Dysphagia
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ORIGINAL ARTICLE

Dysphagia in Progressive Supranuclear Palsy

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Abstract
Progressive supranuclear palsy (PSP) is the most common Parkinson-Plus syndrome and is associated with early onset of dysphagia relative to Parkinson Disease. The current study contributes to the growing understanding of swallowing dysfunction in PSP by describing oropharyngeal swallowing characteristics in a large prospective cohort of participants with PSP employing a nationally standardized videofluoroscopy protocol and a disease severity scale developed expressly for PSP. Participants were 51 adults diagnosed with PSP. Each participant underwent a clinical interview and standardized videofluorographic assessment. Swallowing function was characterized with the Modified Barium Swallow Impairment Scale (MBSImP) and Penetration–Aspiration Scale (PAS). Variables of interest were participant-reported difficulties with liquids and/or solids; overall impression score for each of the 17 individual MBSImP components, as well as Oral Total Sum and Pharyngeal Total Sum; and PAS. Data were described with median interquartile range, counts, and proportions. Spearman's rank correlations were calculated between MBSImP scores and participant-reported indices, FOIS, and PSP Rating Scale. Approximately two-thirds of participants reported difficulties with liquids, solids, or both, although fewer than 15% reported modifying consistencies. Videofluorographic findings included predominant oral phase impairments, including back and forth rocking motion of the tongue, delayed initiation of the pharyngeal swallow, and oral residue. Pharyngeal phase impairments were relatively infrequent and comparatively mild, with the exception of reduced tongue base retraction contributing to pharyngeal residue, and mildly disrupted laryngeal vestibule closure. Disease severity correlated significantly with oral ($r = .0.42$, $p = .0.002$) and pharyngeal ($r = 0.41$, $p = .0.003$) total sum scores as well as with the oral phase components of oral transport ($r = .0.33$, $p = .0.02$) and initiation of the pharyngeal swallow ($r = .0.38$, $p = .0.007$), and PAS for thin liquids ($r = .0.44$, $p = .0.001$). The PSP Rating Scale was not more strongly correlated with swallowing impairment than has been reported for other disease severity rating scales. Dysphagia is a common complaint of patients with PSP. The current findings corroborate and expand upon those reported in the literature, detailing relatively more frequent and more severe oral phase impairments and relatively spared hyolaryngeal excursion. Further research is needed to characterize the progression of dysphagia in PSP and to determine whether dysphagia varies in character or in rate of progression across variants of PSP.

Keywords Deglutition · Deglutition disorders · Swallowing · Swallowing disorders · Progressive supranuclear palsy · Parkinsonism · Modified barium swallow

Introduction

Progressive supranuclear palsy (PSP) is the most common Parkinson-Plus syndrome and is characterized by vertical supranuclear gaze palsy and postural instability, typically with falls occurring within the first year of symptom onset. The presenting symptoms vary and include features of parkinsonism such as rigidity and akinesia, as well as cognitive dysfunction, dysphagia, and dysarthria. Bulbar symptoms are common in PSP, and typically occur earlier in the disease

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course compared to Parkinson's disease [1, 2], although this has not been a universal finding [3].

Oropharyngeal dysphagia is a well-documented symptom of PSP [3–7], typically noted 3–4 years after disease onset, with earlier onset associated with shorter survival [1]. Studies of swallowing function in PSP have included reports of patient complaints [3, 8, 9] as well as radiographic [3, 9, 10], electrophysiologic [8], endoscopic [11], and ultrasound [9] findings. Commonly reported abnormalities include disrupted lingual movements affecting oral containment [6, 9–11], mastication [3, 10], bolus propulsion [6, 9, 10], and oral clearance [6, 12]; delayed onset of the pharyngeal swallow [3, 6, 8, 11]; disrupted velar function [6, 9, 10]; pharyngeal residue [3, 6, 9–11]; penetration and aspiration [3, 6, 10–12]; disrupted function of the upper esophageal sphincter [3, 8, 9]; and esophageal dysmotility [3, 9, 10, 13].

These prior reports emphasized clinical signs of disrupted bolus transport and airway incursion, with biomechanical and physiologic details presented for tongue, palate, and epiglottic movements. Incomplete descriptions are available for several biomechanical features known to contribute to pharyngeal clearance and airway protection, including hyolaryngeal excursion and pharyngeal peristalsis. Such information would lend important insight into the source of swallowing impairment in PSP, as well as direct efforts for clinical trials targeting behavioral interventions such as swallowing maneuvers and strengthening exercises. The current study employed the Modified Barium Swallow Impairment Profile (MBSImP) [14], a standardized protocol for administration and scoring of videofluorographic swallowing assessment that addresses parameters previously noted to be impaired in PSP as well as several measures not yet attributed to this condition, including laryngeal elevation, anterior hyoid excursion, and pharyngeal peristalsis. Moreover, the MBSImP offers summary scores to capture overall severity of oral and pharyngeal impairments, which may be sensitive to disease progression.

Previous descriptions of swallowing dysfunction in PSP have characterized disease severity using tools developed to feature the severity of Parkinson Disease, such as Hoehn and Yahr [3–5, 15] and the Unified Parkinson Disease Rating Scale (UPDRS) [5, 8, 10, 11, 16]. Although useful for characterizing impairments noted in Parkinson Disease specifically, they fail to sample the full range of symptoms of PSP. The current study is the first to employ the PSP Rating Scale [17] to examine the relationship between features of dysphagia and PSP disease severity. We hypothesize that severity of both oral and pharyngeal impairments will be associated with disease severity.

Methods

Participants

Participants included 51 individuals diagnosed with probable or possible PSP who are enrolled in an NIH-funded study (R01 NS089757). The diagnosis of PSP was established by neurologic examination based on international consensus criteria for diagnosing PSP variants [18]. Table 1 lists participant characteristics including PSP Rating Scale scores and PSP variant.[17].

Table 1 Participant characteristics

	N = 51
No. of females, *n* (%)	25 (49%)
Education (years)	15 [12, 16] (8, 20)
Age at onset (years)	66 [62, 71] (49, 80)
Age at clinical exam (years)	71 [65, 75] (54, 86)
Disease duration (years)	4 [2, 5] (1, 10)
PSP Rating Scale[a] (PSP-RS)	42 [35, 50] (17, 67)
Diagnosis[b], *n* (%)	
Probable PSP-RS (Richardson syndrome): vertical supranuclear gaze palsy and postural instability	33 (65%)
Probable PSP-P (Parkinsonism): asymmetric onset of tremor, moderate initial response to levodopa, slower rate of progression	8 (16%)
Probable PSP-PAGF (Pure Akinesia Gait Freezing): Start hesitation and freezing of gait, speech, and/or writing	4 (8%)
Possible PSP-SL (speech and language): apraxia of speech, agrammatic aphasia	3 (6%)
Possible PSP-CBS (corticobasal syndrome): limb apraxia, alien limb, limb rigidity, akinesia, myoclonus, cortical sensory loss	3 (6%)

Data shown are *n* (%) or median [IQR] (range)

[a]Golbe and Ohman-Strickland [17]

[b]Adapted from Hoglinger et al. [18]

Procedures

Each participant completed a standardized clinical interview regarding their swallowing concerns. Questions specifically targeted whether there were difficulties swallowing liquids and solids and, if challenges were reported, whether the participant was modifying the consistency of their diet in response [3, 6, 8, 9]. The clinical interview included the participant as well as any family members or care providers who accompanied the participant.

The videofluorographic swallowing assessment was conducted in accordance with the MBSImP protocol [14]. Consistencies presented included thin and nectar thick barium (liquid consistencies), E–Z paste (puree consistency) (Varibar) and Lorna Doone shortbread cookie with paste coating (solid consistency). The volume and order of recorded swallows are listed in Table 2. The MBSImP yields ratings for 17 individual components of swallowing function as well as Total Sum scores for Oral and Pharyngeal components. The Penetration–Aspiration Scale (PAS) [19] was also scored for each consistency. Finally, ratings on the Functional Oral Intake Scale (FOIS) [20] were collected to characterize the degree to which, based on the exam findings, the clinician recommended modifying the oral diet or the degree to which non-oral nutrition was recommended. Scoring of the MBSImP, PAS, and FOIS was completed by the first author. Recordings from 10% of the participants were reviewed by the third author for MBSImP and PAS ratings for the purpose of reliability assessment. Both raters are MBSImP certified. Inter-rater Spearman's rank correlation for ratings on individual components of the MBSImP was 0.7348 ($p < 0.001$), with 70% of ratings being in full agreement and an additional 11% of ratings being within one unit on the severity scale. Spearman's rank correlation for PAS ratings was 0.873 ($p < 0.0001$).

Data Analysis

Characteristics of participants were described with median, interquartile range (IQR), range, counts, and proportions. Spearman's rank correlations were calculated between MBSImP scores and participant-reported indices, FOIS, and PSP Rating Scale. The measures of pharyngeal stripping wave and pharyngeal contraction, and PAS for puree and solid consistencies were excluded from correlational analysis because of zero-clustered data. Confidence interval of a Spearman's rank correlation was computed via the bootstrap method. The statistical analyses were performed using R version 3.4.4.

Results

Participant-Reported Difficulties

Approximately two-thirds of the participants reported difficulty swallowing liquids, solids, or both (Table 3). Seven participants (14%) reported modifying liquid consistencies

Table 3 Patient-reported difficulty with liquid and solid consistencies

	N (%age)
Difficulty with liquids	
No difficulty	17 (34%)
Reported difficulty but does not modify consistency	26 (52%)
Modifies consistency	7 (14%)
Difficulty with solids	
No difficulty	19 (38%)
Reported difficulty but does not modify consistency	27 (54%)
Modifies consistency	4 (8%)

Table 2 Videofluoroscopic swallowing examination protocol

	Volume	Modality
Lateral view		
Thin liquid (not included in OI score)	5 ml	Teaspoon
Thin liquid	5 ml	Teaspoon
Thin liquid	Self-selected	Single cup sip
Thin liquid	Self-selected	Sequential swallow
Nectar liquid	5 ml	Teaspoon
Nectar liquid	Self-selected	Single cup sip
Nectar liquid	Self-selected	Sequential swallow
Pudding	5 ml	Teaspoon
Solid	½ Shortbread cookie coated with 3 ml pudding consistency	
Anterior–posterior view		
Nectar liquid	5 ml	Teaspoon
Pudding	5 ml	Teaspoon

ID	Diagnosis	Age onset	Age exam	Duration
1	PSP-RS	68	69	1.0
2	PSP-RS	57	59	2.0
3	PSP-RS	62	64	2.0
4	PSP-RS	63	65	2.0
5	PSP-RS	63	65	2.0
6	PSP-RS	63	65	2.0
7	PSP-RS	68	70	2.0
8	PSP-RS	69	71	2.0
9	PSP-RS	69	71	2.0
10	PSP-RS	72	74	2.0
11	PSP-RS	74	76	2.0
12	PSP-PGF	75	77	2.0
13	PSP-PGF	80	82	2.0
14	PSP-RS	52	54	2.5
15	PSP-CBS	56	59	3.0
16	PSP-RS	59	62	3.0
17	PSP-RS	62	65	3.0
18	PSP-PGF	63	66	3.0
19	PSP-RS	63	66	3.0
20	PSP-RS	65	68	3.0
21	PSP-RS	70	73	3.0
22	PSP-PGF	71	74	3.0
23	PSP-RS	72	75	3.0
24	PSP-RS	55	58	3.5
25	PSP-CBS	61	65	4.0
26	PSP-RS	61	65	4.0
27	PSP-RS	64	68	4.0
28	PSP-RS	65	69	4.0
29	PSP-RS	67	71	4.0
30	PSP-RS	69	73	4.0
31	PSP-CBS	71	75	4.0
32	PSP-RS	71	75	4.0
33	PSP-RS	72	76	4.0
34	PSP-P	74	78	4.0
35	PSP-P	49	54	5.0
36	PSP-RS	54	59	5.0
37	PSP-RS	58	63	5.0
38	PSP-RS	65	70	5.0
39	PSP-RS	66	71	5.0
40	PSP-RS	67	72	5.0
41	PSP-P	71	76	5.0
42	PSP-P	67	73	6.0
43	PSP-RS	60	67	7.0
44	PSP-SL	77	85	7.0
45	PSP-P	79	86	7.0
46	PSP-SL	64	72	8.0
47	PSP-P	69	77	8.0
48	PSP-RS	69	77	8.0
49	PSP-SL	71	79	8.0
50	PSP-P	53	63	10.0
51	PSP-P	61	71	10.0

◂**Fig. 1** MBSImP ratings for individual participants organized by disease duration. Age, age of onset, and disease duration are listed in years. The far-right graph depicts the severity rating for each MBSImP component. A higher score, depicted with a darker shade, reflects greater impairment

and four participants (8%) reported modifying solid consistencies.

Videofluorographic Findings

Figure 1 displays the MBSImP profile for each of the participants sorted by disease duration; Figs. 2, 3 and 4 illustrate summary data for oral, pharyngeal, and esophageal domains, respectively. The median Oral Total Sum Score (OTSS) was 8 out of a maximum possible 17 (Fig. 2). Lip closure was unimpaired for 69% of the participants. When disruption of lip closure was present, it was limited to interlabial escape for 11 of the 16 participants. A single participant experienced escape of liquid that flowed beyond the chin, warranting the most severe rating for that item. Tongue control during bolus hold was unimpaired for 67% of participants. Loss of the bolus posteriorly was observed for fewer than 20% of the participants. Slowed but ultimately complete mastication of a solid bolus (bolus preparation) was observed in 59% of participants; another 24% exhibited normal mastication.

Disruption of bolus transport/lingual motion was frequently observed (76% of participants), with the most common pattern characterized by disorganized back and forth motion of the tongue (displayed by 35% of participants). All participants but one exhibited oral residue, with most participants (84%) demonstrating moderate oral residue. Initiation of the pharyngeal swallow was delayed in 76% of participants. For 43% of participants, the delay involved the bolus reaching the level of the pyriforms before initiation of the pharyngeal response.

The median Pharyngeal Total Sum Score (PTSS) was 2 out of a maximum possible 31. Figure 3 illustrates that pharyngeal phase components were frequently normal, as indicated by a score of 0. In particular, soft palate elevation, laryngeal elevation, anterior hyoid excursion, epiglottic movement, pharyngeal stripping wave, pharyngeal contraction, and pharyngo-esophageal (PE) segment opening were each impaired in fewer than 12% of the participants.

Impaired laryngeal vestibule closure, most often characterized by a narrow column of contrast in the laryngeal vestibule, was observed in 53% of the participants. Reduced tongue base retraction, resulting in observable contrast or air between the tongue base and the pharyngeal wall was evident in 45% of the participants and was a contributing factor to pharyngeal residue, which was the most frequently observed pharyngeal impairment (present for 73% of participants).

PE segment opening was normal in 90% of participants. Of the five participants for whom any abnormality of PE segment function was observed, only one demonstrated minimal distension or duration with marked obstruction of flow.

Esophageal clearance was normal for 55% of the participants (Fig. 4). With the exception of one participant who experienced retrograde flow, the remaining participants with esophageal findings demonstrated esophageal retention without retrograde flow.

PAS scores are summarized in Fig. 5. The median score on the PAS for thin liquid was 2, with 45% of the participants demonstrating no airway incursion with any of the consistencies presented (PAS score of 1). Aspiration of thin liquids without effort to eject (PAS score of 8; silent aspiration) was observed in five participants. The median PAS for nectar liquid was 1, with aspiration (PAS score of 6 or higher) observed for only two participants. All puree and solid consistency swallows were rated as a PAS of 1, with the exception of a single participant who demonstrated PAS of 2 for puree.

The most common FOIS rating was 7 (total oral intake with no restrictions, 65%), followed by 5 (total oral intake of multiple consistencies requiring special preparation, 31%) and 6 (total oral diet with multiple consistencies without special preparation, but with specific food limitations, 4%). No participants were recommended for non-oral nutrition based on the examination findings.

Correlational Analyses

Pairwise scatter plots among OTSS, PTSS, and the PSP Rating Scale [21] illustrate weak but statistically significant correlations (Fig. 6). The correlation between OTSS and PTSS ($r=0.25$, $p=0.08$) is weaker than the correlation between the PSP Rating Scale and Oral ($r=0.42$, $p=0.002$) and Pharyngeal ($r=0.38$, $p=0.007$) Sum scores. Figure 7 depicts the Spearman's correlations between each of the videofluorographic metrics (MBSImP components, Total Sum scores and PAS), with the participant-reported difficulties and disease severity (PSP Rating Scale). Participant-reported difficulties with liquids correlated significantly with PTSS [$r=0.33$ ($p=0.03$)] and PAS for thin liquids [$r=0.36$ ($p=0.02$)]. Participant-reported difficulties with solids correlated significantly with bolus transport impairments [$r=0.33$ ($p=0.04$)]. Disease severity (PSP Rating Scale) correlated significantly with both OTSS [$r=0.42$ ($p=0.009$)] and PTSS [$r=0.41$ ($p=0.01$)], as well as the individual components of bolus transport [$r=0.33$ ($p=0.04$)] and initiation of the pharyngeal swallow [$r=0.38$ ($p=0.03$). Finally, disease severity correlated significantly with PAS for thin liquids [$r=0.44$ ($p=0.005$)].

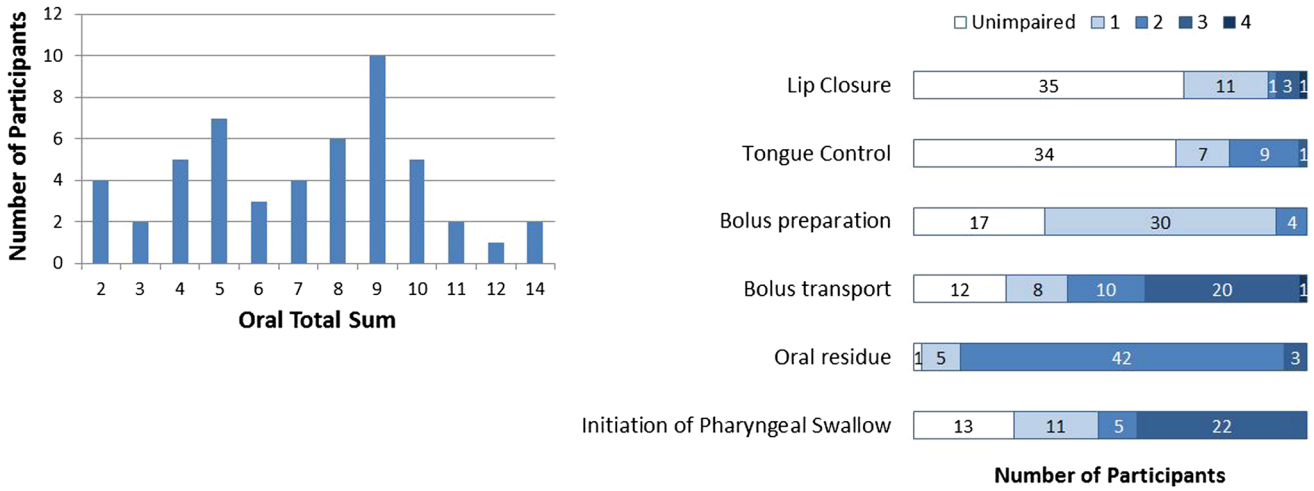

Fig. 2 MBSImP oral domain components. The far-left graph depicts the number of participants exhibiting each of the Oral Total Sum Scores displayed. The far-right graph depicts the number of participants exhibiting each level of severity for each component. A higher score, depicted with a darker shade, reflects greater impairment

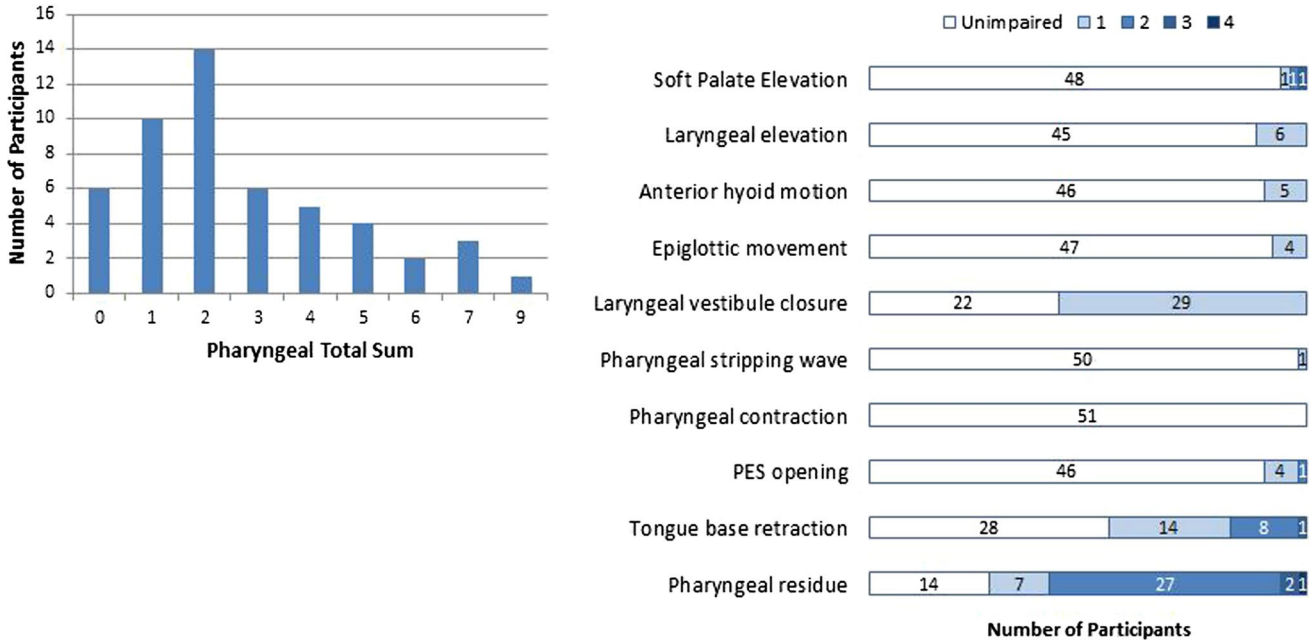

Fig. 3 MBSImP pharyngeal domain components. The far-left graph depicts the number of participants exhibiting each of the Pharyngeal Total Sum Scores displayed. The far-right graph depicts the number of participants exhibiting each level of severity for each component. A higher score, depicted with a darker shade, reflects greater impairment

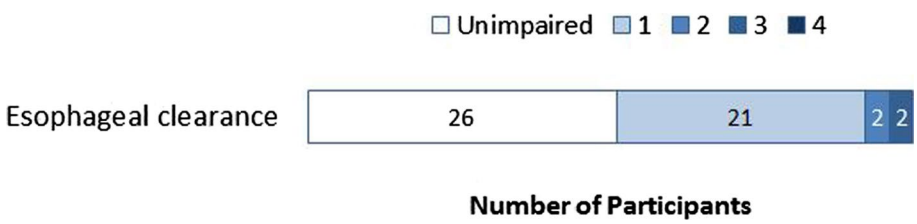

Fig. 4 MBSImP esophageal clearance. The graph depicts the number of participants exhibiting each level of severity. A higher score, depicted with a darker shade, reflects greater impairment

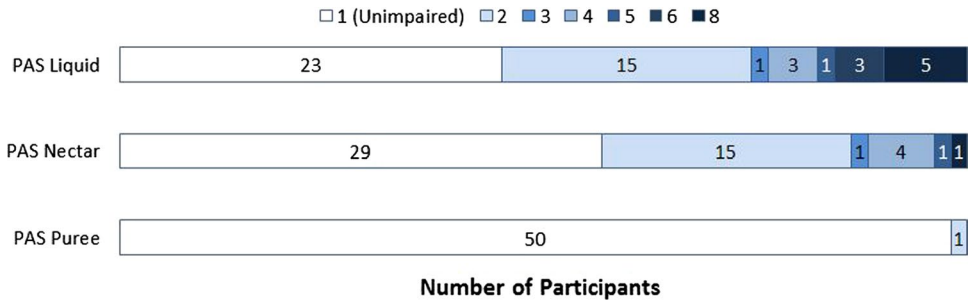

Fig. 5 Penetration Aspiration Scale: highest rating from thin liquid, nectar liquid, and puree consistencies. The graph displays the number of participants rated with the designated score. A higher score reflects greater impairment

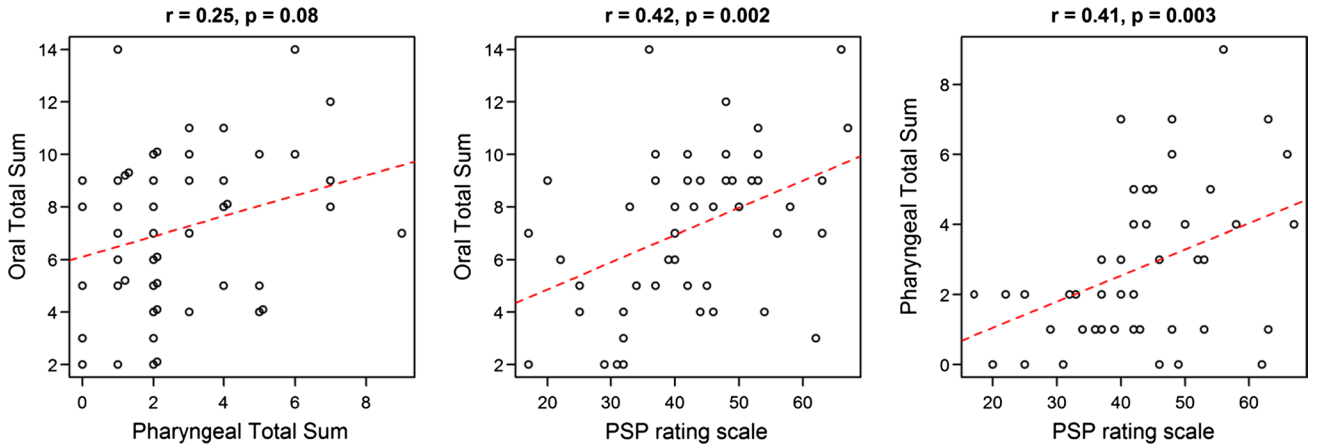

Fig. 6 Scatterplots between Oral Total Sum, Pharyngeal Total Sum, and PSP rating score with Spearman's correlations

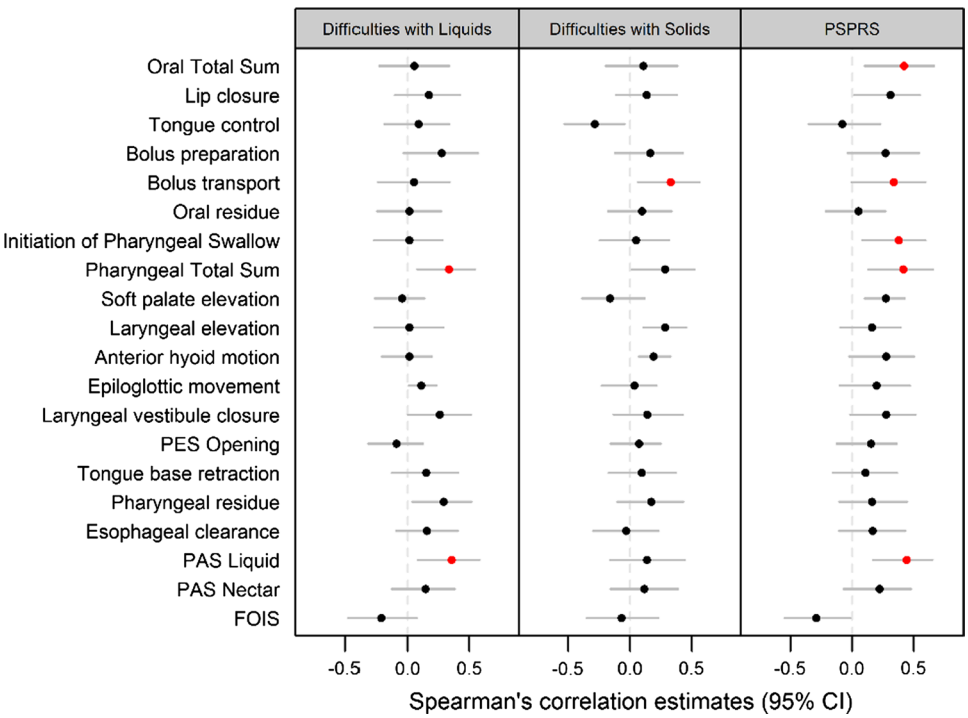

Fig. 7 Plots of Spearman's correlations. Red dots represent those with FDR q value < 0.05

Discussion

Progressive supranuclear palsy is a Parkinson's plus syndrome with well-documented impact on oropharyngeal swallowing. The current study is the largest to describe swallowing function in PSP, the first to report MBSImP profiles in PSP, and the first to employ the PSP Rating Scale to examine the relationship between features of dysphagia and PSP disease severity.

Participants commonly reported difficulties swallowing both liquids and solids, although few reported modifying consistencies in response to the difficulties they experience. Participants' complaints typically aligned with observed clinical impairments, with moderate and statistically significant correlations noted between complaints of difficulties swallowing liquids and PTSS and PAS for thin liquids, as well as between difficulties swallowing solids and impairments of bolus transport.

Videofluorographic analyses revealed oral phase components were more severely impaired than were pharyngeal phase components. This finding is in keeping with a number of previous investigations [3, 6, 10] reporting impaired lingual function as evidenced by back and forth rocking of the tongue during attempts to propel the bolus posteriorly, reduced tongue base retraction, and delayed onset of the pharyngeal swallow. Indeed, the penetration and aspiration observed in the current study consistently occurred before the swallow as a result of delayed pharyngeal response.

Impaired velar functioning is reported in previous studies [6, 9, 10], but not commonly observed in the current study. This could partially reflect the rating scales used. Early studies attributed posterior spillage to inappropriate velar retraction and elevation during the oral phase of the swallow. The MBSImP emphasizes the role of the tongue, rather than the palate, in creating the posterior oral seal. Ratings of palatal function are instead made based on integrity of elevation, which is a pharyngeal phase component. In the current study, posterior spillage was observed for roughly one-third of the participants. Future studies could further elucidate the contribution of abnormal palatal movements to incomplete posterior seal by including additional description of linguapalatal biomechanics when posterior spillage is observed.

Relative sparing of laryngeal elevation and anterior hyoid excursion is particularly notable as these two biomechanical phenomena contribute to airway protection by displacing the entrance to the laryngeal vestibule and by promoting epiglottic inversion. Additionally, adequate hyolaryngeal excursion promotes clearance of the bolus from the pharynx by allowing the valleculae to empty and by contributing to the opening of the pharyngo-esophageal segment. The consistent sparing of hyolaryngeal biomechanics, as well as the typically intact pharyngeal stripping wave, likely accounts for the generally mild pharyngeal residue and infrequent aspiration that was observed in this clinical cohort.

While pharyngeal phase impairments were typically mild relative to oral phase impairments, disease severity (PSP Rating Scale) did correlate significantly with PTSS. Indeed, PTSS and PAS were correlated significantly only with disease severity. This might suggest that emergence of clinically significant pharyngeal dysphagia is a landmark in PSP progression and provides support for close monitoring of swallowing function in patients with PSP.

The current study is the first to examine the relationship of swallowing function to disease severity as measured by the PSP Rating Scale. Observed Spearman correlations were 0.42 and 0.41 for OTSS and PTSS, respectively. These findings are comparable to correlations between dysphagia severity score and Hoehn and Yahr [15] (0.472) and the UPDRS [16] (0.474) reported by Warneke et al. [11], but lower than those reported by Alfonsi et al. [8], who noted correlations ranging from 0.64 to 0.793 between UPDRS and duration of key electrophysiologic swallowing events. Taken together, the current findings did not suggest a clear advantage of PSP Rating Scale over alternate indices of disease severity previously employed for the study of dysphagia in PSP.

Neuroanatomic Explanation for Findings

It is well established that both brainstem and supranuclear control centers are active during deglutition. Traditional descriptions characterize the oral phase of the swallow as under voluntary control, with greater influence of supranuclear control centers [22] and the pharyngeal and esophageal phases driven by a medullary central pattern generator (CPG) [23]. Early studies of swallow function in parkinsonian syndromes hypothesized that degeneration of the cholinergic neurons of the pedunculopontine tegmental nucleus (PPTN) contributed to dysphagia [8, 11]. The PPTN has connections with the basal ganglia and projects to the nucleus tractus solitarius, part of the swallowing CPG. The early progression of dysphagia in PSP is attributed to degeneration of the neurons in the CPG itself [24], pseudobulbar impairments [10], neck hyperextension [6], and deficits in attention and executive function [11]. In spite of the considerable speculation about the source of dysphagia in PSP, no studies to date have systematically examined neuroanatomical correlates to the presence, nature, and/or severity of dysphagia in PSP. Future work will address this question.

Clinical Implications

The pattern of swallowing impairment observed in PSP can inform not only symptom-focused compensatory strategies but also impairment-focused rehabilitative and/or prophylactic exercises. Given the disproportionate oral phase impairment with notable disruption of lingual functions, lingual strengthening may be a reasonable intervention [25]. Exercises targeting hyolaryngeal elevation in relative isolation would not be of highest priority. However, expiratory muscle-strengthening exercises that might have more immediate relevance for addressing non-swallowing impairments of dysarthria and/or dyspnea [26] may have prophylactic benefit for the submental musculature contributing to hyolaryngeal excursion [27]. Each of these hypotheses requires empirical assessment with appropriate experimental designs.

Limitations to the Current Study

A definitive diagnosis of PSP is established by neuropathology only. Thus, while participants met the highest criteria established for the variants ascribed, diagnosis of PSP will not be confirmed until autopsy. The current study, while the largest cohort of PSP participants for whom swallowing function is detailed, was heavily weighted with participants exhibiting the Richardson Syndrome variant. The small sample of participants in several variants precluded between-group analyses across variants. Given the evidence for involvement of distinct neuroanatomic landmarks across PSP variants [28–30], our future research will seek to detail differences in the nature and severity of swallowing disruption across variants.

The study utilized perceptual judgments for rating parameters of the swallow. This offered clear advantage for translation given the wide employment of the MBSImP. Nonetheless, other kinematic, temporal, or electrophysiologic measures may prove more useful for identifying subtle impairments, particularly in the pharyngeal phase of swallowing where temporal features are not captured by the MBSImP.

Conclusions

The current study describes swallowing function in a large cohort of individuals diagnosed with PSP. Consistent with previous studies, we demonstrated that oral phase impairments, including back and forth rocking of the tongue and delayed initiation of the swallow response are much more prominent than pharyngeal phase impairments, particularly disruption of hyolaryngeal elevation. Vallecular residue, although usually mild, was evident in the majority of participants and was attributable to reduced tongue base retraction in nearly half of the participants. Further research is needed to determine whether swallowing function varies either in character or in rate of progression across variants of PSP.

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Compliance with Ethical Standards

Conflict of interest Authors Clark, Stierwalt, Tosakulwong, Botha and Ali have no conflicts to disclose. Drs. Botha, Whitwell & Josephs receive research support from the National Institute of Health.

Ethical Approval All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

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