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Dysphagia in progressive supranuclear palsy: A scoping review

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ABSTRACT

Introduction: One of the most prevalent types of atypical parkinsonian syndrome is progressive supranuclear palsy (PSP). PSP is associated with early onset of dysphagia which can result in malnutrition, dehydration, and aspiration pneumonia, affecting quality of life and increasing mortality rate. To date, research describing dysphagia in PSP and its impact is scant.

Methods: The objective of this scoping review is to determine the characteristics of dysphagia in PSP, differences in dysphagia presentation according to PSP subtype, principal methods used for identifying and diagnosing dysphagia and the impact dysphagia has on quality of life in individuals with PSP. This review was conducted in accordance with the JBI methodology. Six electronic databases were searched.

Results: Of the 20 studies included, the most frequently reported characteristics of dysphagia were oral preparatory and oral phase difficulties. A variety of methods were used to identify and diagnose dysphagia including instrumental assessment (65%), patient reported scales (45%) and clinical swallow evaluation (20%). The most used instrumental assessment was videofluoroscopy (46%). Limited data was available describing characteristics of dysphagia according to the subtype of PSP. The impact that dysphagia has on quality of life was assessed in only one study.

Conclusion: A range of assessment methods are used to identify and diagnose dysphagia in patients with PSP. Further research is needed to investigate if particular characteristics are associated with certain PSP subtypes. Future studies should also measure the impact that dysphagia has on quality of life in this population.

1. Introduction

Atypical parkinsonian syndromes (APS) encompass a collective of rare neurodegenerative diseases and are characterized by rapid disease progression and decreased life expectancy [1]. APS can be particularly debilitating and yet research in this area is limited in comparison to other neurodegenerative conditions of similar prevalence. One of the most common APS is progressive supranuclear palsy (PSP).

PSP was initially characterized by postural instability, supranuclear vertical gaze palsy leading to impaired vision, dementia, dysarthria, and other features including swallowing disorders (dysphagia) [2,3]. Two main subtypes of PSP have primarily been described; Richardson syndrome (PSP-RS) and PSP-parkinsonism (PSP-P) [4]. PSP-RS is the most common type of PSP and is characterized by early onset of postural instability and falls, vertical supranuclear gaze palsy, and cognitive dysfunction. PSP-RS is associated with faster disease progression and a shorter survival time [2]. In contrast, PSP-P is characterized by asymmetric onset, tremor, and a moderate, if unsustained, response to levodopa. PSP-P is often misdiagnosed as idiopathic Parkinson's disease (IPD) because of overlapping characteristics. The prevalence of the classic Richardson syndrome is estimated to be up to 6.4/100,000 [5,6]. However, more recent research suggests that this figure may be higher, with average age of onset in the mid-60s and a disease duration from diagnosis to death of approximately six years [7].

Several other variants of PSP have been described by the Movement Disorder Society (MDS) based on initial clinical presentations [8]. The MDS outlined criteria aimed at optimizing early, sensitive, and specific clinical diagnosis of PSP based on current evidence [8]. As well as PSP-RS and PSP-P, other subtypes include, PSP-OM (ocular motor dysfunction), PSP-PI (postural instability), PSP-F (frontal lobe cognitive or

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behavioral presentations), PSP-PGF (progressive gait freezing), PSP-CBS (corticobasal syndrome) and PSP-SL (speech/language disorders) [8]. As the disease progresses, subtypes may evolve into PSP-RS.

Dysphagia is common in patients with PSP and its subtypes, yet it is underdiagnosed and methods of assessing and determining the presence of dysphagia vary [9]. Dysphagia tends to present at the early stages of the condition and is typically assessed through completion of a clinical swallow evaluation and instrumental assessments such as videofluoroscopy (VFSS) and fiberoptic endoscopic evaluation of swallowing (FEES) [10]. Dysphagia can result in clinical complications, including malnutrition, dehydration, and aspiration pneumonia, potentially affecting quality of life (QOL) and eventually increasing the mortality rate. Research investigating the impact dysphagia has on QOL in individuals with PSP is limited, despite dysphagia being identified as having a significant effect on QOL in other neurological conditions such as IPD [11]. Aspiration pneumonia secondary to dysphagia is a major risk in advanced PSP and is the principal cause of death [12].

A review of the literature in relation to the characteristics of dysphagia in PSP and variations in dysphagia presentation according to PSP subtypes is warranted, as the consequences of dysphagia can be significant and evidence in this field is limited. Furthermore, establishing the key methods used to identify dysphagia and the impact dysphagia has on QOL in this population will inform future research which may aid in earlier diagnosis and management of PSP and its subtypes. An improved understanding of the nature, diagnostic methods, and impact that dysphagia has on this population may reduce adverse clinical and QOL outcomes.

The objective of this scoping review is to determine the principal methods used for identifying and diagnosing dysphagia, the characteristics of dysphagia, and the impact dysphagia has on QOL in individuals with PSP. It is also hypothesized that there may be differences in dysphagia presentation according to the subtype of PSP.

Review questions:

- What are the key methods for identifying and diagnosing dysphagia in this population?
- What are the characteristics of dysphagia in people with PSP including all PSP subtypes?
- Are there differences in dysphagia presentation according to the subtype of PSP?
- What impact does dysphagia have on quality of life in people with PSP?

2. Methods

This scoping review was conducted in accordance with JBI methodology for scoping reviews [13]. The Preferred Reporting Items for Systematic Reviews and Meta Analysis extension for Scoping Reviews (PRISMA-ScR) were used to guide the review [13] (supplemental material 1). A preliminary search of MEDLINE, PROSPERO, the Cochrane Database of Systematic Reviews and JBI Evidence Synthesis was conducted and no current or in-progress systematic reviews or scoping reviews on the topic were identified. A research protocol was published in December 2022 [14].

2.1. Eligibility criteria

Studies which included participants with a clinical diagnosis of PSP established by a neurologic exam and dysphagia were included. Studies were excluded if participants had a comorbidity, which could independently cause dysphagia. Studies published since database inception to July 2023, based in any country, in any setting and in any language were included. We considered both experimental and quasi experimental study designs including randomized controlled trials, non-randomized controlled trials, before and after studies and interrupted time-series studies. In addition, analytical observational studies including prospective and retrospective cohort studies, case-control studies and analytical cross-sectional studies were considered for inclusion. We also considered descriptive observational study designs including case series, individual case reports and descriptive cross-sectional studies for inclusion.

In addition, systematic reviews were also considered. Grey literature including conference posters and presentations were included if sufficient detail was available. Editorials, expert opinions and secondary research were excluded.

2.2. Search strategy

The search strategy aimed to identify both published and unpublished studies. A preliminary search of PubMed and CINAHL was undertaken to identify articles on the topic. The text words contained in the titles and abstracts of relevant articles, and the index terms used to describe the articles were used to develop a full search strategy with the assistance of an expert subject librarian (supplemental material 2). The search strategy, including all identified keywords and index terms, was adapted for each included database and/or information source. Reference lists of all included sources of evidence were screened for additional studies. Databases searched included PubMed, EMBASE, CINAHL, Web of Science Core Collection and PsycINFO. Sources of unpublished studies/grey literature searched included ProQuest Dissertation and Theses Global.

2.3. Selection of studies

All identified citations were uploaded into an online platform Covidence (https://www.covidence.org) and duplicates were removed. Following a pilot test, titles and abstracts were screened by two independent reviewers (EF, JG). Potentially relevant sources were retrieved in full, and their citation details imported. The full texts of selected citations were reviewed in detail against the eligibility criteria by the two independent reviewers (EF, JG). If studies were unobtainable or additional data were required, authors were contacted only if studies had been completed in the previous five years. If there was no contact from authors after a two-week period, studies were excluded. Reasons for exclusion at full text stage were recorded. Any disagreements were resolved through discussion, or with additional reviewers (MW, JR). Articles in languages other than English were translated using translation software (https://www.deepl.com).

2.4. Data charting process

Data were extracted by two reviewers (EF, JG) using a data extraction tool developed in Covidence. The tool was piloted independently on three included studies (EF, JG). Following this, minor modifications were made. The data extracted included specific details about the participants, concept, context, study methods and key findings relevant to the review questions. If a study had data for more than one timepoint, data at the first visit only was used to allow for comparison with other studies in the review which provided data at one timepoint only, typically that at the first clinical visit.

2.5. Data analysis and presentation

Data relating to dysphagia characteristics were categorized into oral preparatory/oral stage difficulties, pharyngeal stage difficulties and esophageal stage difficulties. Methods of dysphagia assessment were categorized into clinical (e.g. clinical swallow evaluation), instrumental assessment methods (e.g. FEES, VFSS, high resolution pharyngeal manometry etc.), swallow screening, and patient reported outcome measures. Validated QOL tools and relevant patient-reported outcome measures were used to describe the impact dysphagia has on QOL in people with PSP. Differences in dysphagia presentation according to the

subtype of PSP were analyzed.

3. Results

3.1. Search results

The search identified 932 study abstracts which were imported to Covidence for screening. 377 duplicates were removed. 555 abstracts were screened, and 313 studies were deemed irrelevant following screening. 242 studies were reviewed at full text stage for potential inclusion. 222 studies were excluded at this point. Interrater agreement during title and abstract screening was 85 % (Cohens's $\kappa = 0.71$) and during full text screening was 96 %, (Cohens's $\kappa = 0.82$), which corresponds to strong and almost perfect agreement, respectively.

The most common reasons for exclusion of studies included inability to obtain full texts or insufficient information on dysphagia presentation. Detailed reasons for exclusion are described in the PRISMA Flow Diagram (Fig. 1). Twenty studies were deemed eligible for inclusion in the review following the study selection process.

3.2. General study characteristics

Table 1 outlines the general characteristics of included studies. Most studies were completed in the USA (6/20, 30 %) [15–20], Italy (6/20, 30%) [21–26] and Germany (4/20, 20%) [27–30]. The remainder of included studies were completed in South Korea (2/20, 10%) [31,32], France (1/20, 5%) [33] and India (1/20, 5%) [34]. The study design most used was case control design (11/20, 55%). A total of 1,012 participants aged between 55–88 years took part in all 20 included studies. Most participants were male (n = 563, 56%) (Table 1). The severity of PSP was rated predominantly using two scales, The Hoehn and Yahr (H&Y) scale [35] (7/20, 35%) and Progressive Supranuclear Palsy Rating Scale (PSPRS) [36] (5/20, 25%). Mean scores are reported as these were the only data published for the majority of included studies [17,25,29]. The mean PSPRS score for participants (n = 679) was 38.61/100 suggesting moderate disability. The mean participant H&Y scale score (n = 198) was 3.3/5 indicating "mild to moderate bilateral involvement, some postural instability but physically independent" [35] (supplemental material 3).

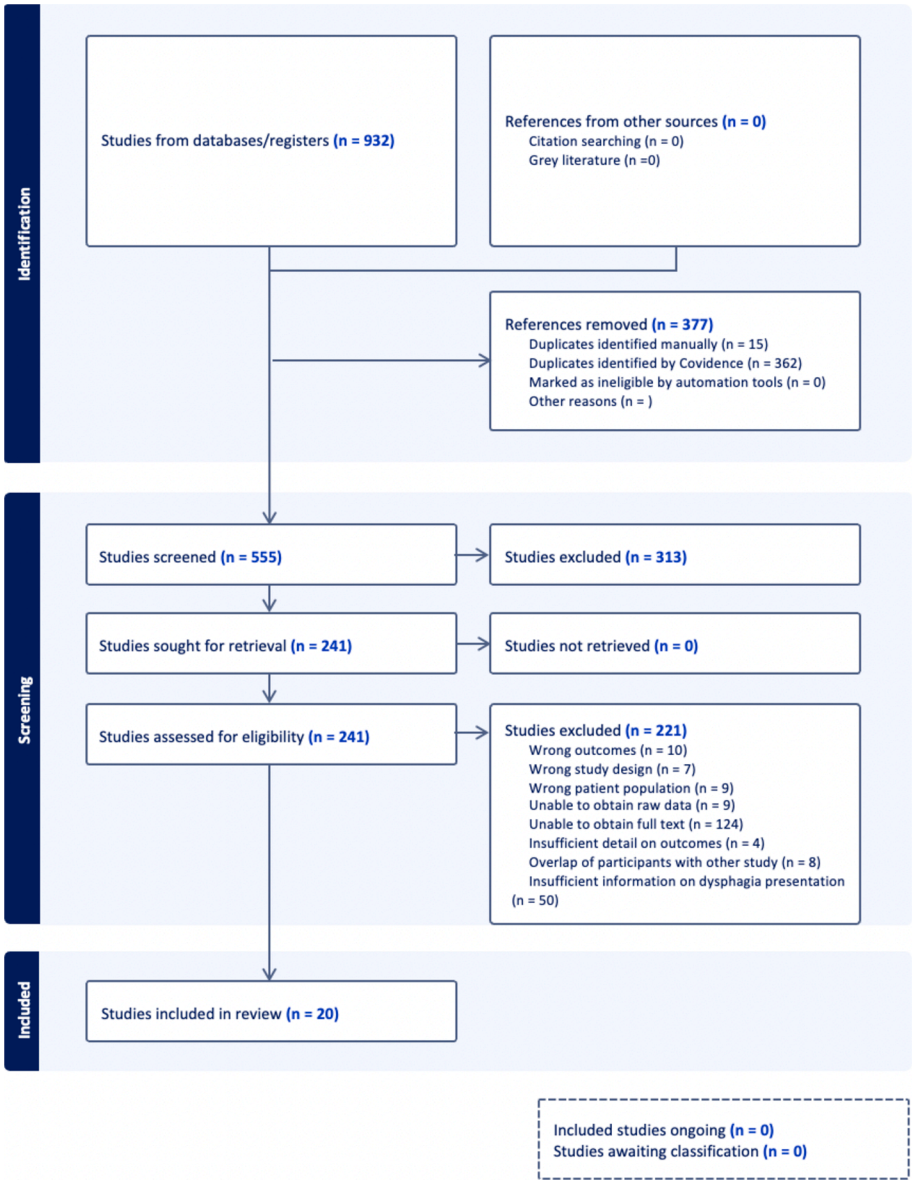

Fig. 1. PRISMA flow diagram illustrating flow of information through the different phases of the scoping review. It maps out the number of identified records, included and excluded, and the reasons for exclusion.

Table 1
General Study Characteristics.

Author, year	Country of origin	Study design	N participants with PSP	Age (years)	Sex	N Disease subtype	Disease duration (years) Unless otherwise specified	Disease severity Mean (range/SD) Unless otherwise specified	Method of swallowing assessment
Alfonsi et al., 2007 [21]	Italy	Case control	9	Mean = 71 (Range = 64–77)	M = 4 F = 5	Not described	Mean = 4 (Range = 3–6)	UPDRS = 41 (Range = 29–59)	• Surface and needle EMG • Dysphagia Severity Score
Alfonsi et al., 2010 [22]	Italy	Case control	9	Mean = 68 (Range = 61–78)	M = 5 F = 4	Not described	Mean = 48.78 months	Not described	• Surface and needle EMG • Dysphagia Severity Score
Beschin et al., 2018 [23]	Italy	Case report	1	60	M = 1	Not described	4	Not described	• MDS-UPDRS • Functional oral intake scale • Swallowing Disturbance Questionnaire
Borders et al., 2023 [15]	USA	Cohort study	24	Mean = 71 (SD = 7.3)	M = 16 F = 8	PSP-RS = 16 PSP-P = 7 PSP-F = 1	Mean = 5.05 (SD = 2.2)	Schwab and England ADL scale = 51.70 (SD = 21.50)	• FEES
Choi et al., 2021 [31]	South Korea	Case control study	123	Mean = 68 (SD = 6.3)	M = 68 F = 55	PSP-RS = 66 PSP-P = 28 PSP-PGF = 29	Mean = 3.7 (SD = 2.6)	H&Y Scale Stage = 3.2 (Range = 2–5)	• PSP Rating Scale Question 13
Clark et al., 2020 [16]	USA	Cross sectional	51	Mean = 68 (Range = 54–86) Median = 71	M = 26 F = 25	PSP-RS = 33 PSP-P = 8 PSP-PGF = 4 PSP-SL = 3 PSP-CBS = 3	Median = 4 (Range = 1–10)	PSPRS Median = 42 (Range = 7–67) IQR: [35, 50]	• VFSS • FOIS • Clinical interview • Patient reported scale
Claus et al., 2018 [27]	Germany	Case control	10	Mean = 72.6 (SD = 3.8)	M = 9 F = 1	PSP-RS = 6 PSP-P = 3 PSP-PGF = 1	Mean = 2.5 (SD = 0.9)	H&Y Scale Stage = 3.3 (SD = 0.8) UPDRS III = 27.0 (SD = 3.0)	• FEES • Esophageal high resolution manometry
Cosentino et al., 2020[a] [24]	Italy	Case control	11	Non dysphagia (n = 1): 67 Dysphagia (n = 10): 70 (SD = 6.2)	M = 9 F = 2	Not described	Mean = 3.8 (SD = 2)	PSPRS total score: Non-dysphagic: 46 Dysphagia:56.2 (SD = 6.5)	• Clinical Assessment • FEES
Golbe et al., 2020 [17]	USA	Cohort	494 in original data for first visit only	Mean = 72 Median = 72 (SD = 746)	M = 249 F = 245	PSP-RS = 465 PSP-PGF = 3 PSP-P = 17 PSP = SL = 3 PSP-C = 1 PSP-CBS = 3 PSP-F = 2	Mean = 45.5 months (SD = 30.9 months)	PSPRS Score = 41.15 (SD = 15.27) Median = 39	• PSP Rating Scale Questions 3 and 13
Han et al., 2023 [32]	South Korea	Case report	1	53	M = 1	Not described	Not described	Not described	• VFSS
Johnston et al., 1997 [18]	USA	Case control	7	Mean = 69.14 (Range = 55–88) Median: 70	M = 5 F = 2	Not described	Mean = 7 (Range = 3–13) Median: 5	H&Y Scale Stage = 4 (Range = 3–5) Median = 4	• VFSS • Standard esophageal manometry • Severity scale for dysphagia
Kaphan et al., 2008 [33]	France	Case report	1	60	F = 1	Not described	Not described	Not described	• Clinical swallow evaluation • VFSS • Esophageal manometry

(*continued on next page*)

Table 1 (*continued*)

Author, year	Country of origin	Study design	N participants with PSP	Age (years)	Sex	N Disease subtype	Disease duration (years) Unless otherwise specified	Disease severity Mean (range/SD) Unless otherwise specified	Method of swallowing assessment
Leopold et al., 1997 [19]	USA	Case control	10	Mean = 71 (Range = 63–83)	M = 7 F = 3	Unclear	Mean = 4.1 (Range = 2–8)	H&Y Scale Stage = 2.7 (Range = 2–5)	• VFSS
Litvan et al., 1997 [20]	USA	Case control	27	Mean = 64.9 (SD = 1.2)	M = 18 F = 9	Not described	Mean = 52 months (SD = 5 months)	H&Y Scale Stage = 3.4 (SD = 0.1)	• National Institutes of Health Speech Pathology swallowing Questionnaire • Oral Motor Scale • Submental Ultrasound • VFSS
Maetzler et al., 2016 [28]	Germany	Case series	3	Mean = 64 (Range = 50–71)	M = 1 F = 2	PSP-P = 3	Mean = 4.3	H&Y Scale Stage = 3 (Range = 2–4)	• CSE • VFSS
Picillo et al., 2020 [25]	Italy	Case control	21	Mean = 67.1 (SD = 6.3)	M = 12 F = 9	PSP-RS = 17 PSP-P = 2 PSP-PGF = 2	Mean = 3.14 (SD = 2)	PSPRS = 39.29 (SD = 15.93) MDS-UPDRS-III: 35.9 (SD = 15.11)	• PSP Rating Scale Question 13
Piot et al., 2020 [29]	Germany	Cross sectional and cohort	164	Mean = 70.4 (Range = 44–85)	M = 100 F = 62[b]	PSP-RS = 107 Variants = 57	Mean = 3.5 (Range = 0–14, SD = 2.5)	PSPRS = 35.4 (Range = 9–75) (SD = 14)	• PSP-CDS • PSP-QOL
Sulena et al., 2017 [34]	India	Case control	25	Not described	M = 19 F = 6	Not described	Mean = 2 5 (SD = 8.3)	Not described	• Interview • Medical records • Detailed examination for swallowing was done which included 3 oz water swallow test
Varanese et al., 2014 [26]	Italy	Case series	3	Mean = 71 (Range = 66–78)	M = 2 F = 1	Not described	Mean = 4.7	Not described	• PSP Rating Scale Question 13
Warnecke et al., 2010 [30]	Germany	Case control	18	Mean = 70 (SD = 9)	M = 11 F = 7	Not described	Mean = 3.47 (SD = 1.89)	H&Y Scale Stage: 3.5 (Range = 2.5–5) UPDRSIII = 37.39 (SD = 13.92)	• FEES

[a] Patients with severe dysphagia were excluded from this study [b] Missing data for two participants.

3.3. Key methods for identifying and diagnosing dysphagia

A range of assessments were used to identify, diagnose, and evaluate dysphagia. These included instrumental assessments (13/20, 65%), patient-reported scales (9/20, 45%), non-instrumental clinical swallow evaluation (4/20, 20%) and swallow screening assessments (1/20, 5%). Eight studies (40%) used more than one assessment type.

3.3.1. Instrumental assessments

Of the studies using instrumental assessment, 6/13 (46%) used VFSS, 4/13 (31%) used FEES. 3/13 (23%) used esophageal manometry; two of these three studies appeared to use standard manometry and one study use high resolution manometry. 2/13 (15%) used electromyography (EMG) and submental ultrasound was used in one study (8%).

3.3.2. Patient reported scales

Of the included studies, just over half (11/20) used a patient reported scale which captured dysphagia severity or characteristics. A range of measures were used including the Dysphagia Severity Score [21,22], the Swallowing Disturbance Questionnaire [37] and the National Institutes of Health Speech Pathology Swallowing Questionnaire [38]. A "self-rated dysphagia scale" was used in one study [18] and Clark and colleagues [16] used a patient-reported scale which assessed patient-reported difficulty with liquid and solid consistencies (1/11). The most frequently used patient reported scale was the PSPRS (5/11) [36]. This scale includes a self-reported scale for solids (Question 3) and a screening element for liquids (Question 13).

3.3.3. Other non-validated clinical assessments

Clinical swallow evaluations (CSE) were used in four studies (20%). A swallow screening tool [39] was used in one study to identify possible dysphagia [34]. A detailed description of the screening tool outlining steps taken when using this test was provided by authors in this study.

3.4. Characteristics of dysphagia

3.4.1. Instrumental assessments

Characteristics of dysphagia are described for studies using instrumental assessments as these are considered to be the most objective measures of swallow function. A wide range of characteristics were assessed over all studies which would not be possible to describe in detail. Characteristics of dysphagia as described using instrumental assessment were considered in this review if over 30 participants were assessed for that specific characteristic. Airway penetration and aspiration were reported for level 0 thin liquids only due to variability in terminology used in included studies for other consistencies. See

supplemental material 4 for a detailed breakdown of characteristics of dysphagia.

The most frequently reported characteristics of dysphagia in included studies which used instrumental assessments were oral preparatory and oral phase difficulties specifically oral residue (51/52, 98%) and bolus transfer/lingual motion difficulties (57/74, 77%) (Fig. 2). The most common pharyngeal phase difficulties reported were pharyngeal residue (48/69, 70%) and delayed initiation of the pharyngeal swallow (73/107, 68%). The esophageal phase of the swallow was less frequently assessed in included studies. Studies which evaluated this phase of the swallow described reflux/gastro esophageal reflux disease (GERD) (20/36, 56%) and esophageal clearance difficulties (25/51, 49%) most frequently (Fig. 1).

3.4.2. Patient-reported scales

Data from the PSPRS were published or obtained from authors for items related to dysphagia in three studies [25,31,17]. One study had PSPRS data for more than one timepoint [17] and as per protocol, data from the first visit only was included.

For Question 13 on PSPRS that related to dysphagia for liquids, Golbe et al [17] found that participants had a median score of 1/4 (range = 0–4) (n = 494), Picillo et al [25] reported a median score of 2/4 (range = 0–3) (n = 21) and Choi et al [31] reported a score median of 0/4 (range = 0–4) (n = 123). Original data from the PSPRS were obtained for two studies [25,17]. This enabled the combination of scores for Question 13 from the PSPRS for three studies [25,31,17]. The median score for Question 13 for all participants (n = 638) in these studies was 0 (range = 0–4) suggesting that most participants did not experience any difficulties swallowing liquids.

Data relating to dysphagia for solids based on Question 3 from the PSPRS were provided in one study [17]. Golbe and colleagues [17] reported a median score of 0 (range = 0–4) (n = 494) indicating that the majority of participants reported "no difficulty with a full range of food textures" [36].

Clark et al [16] used a patient-reported questionnaire which appeared to be developed specifically for this study to rate levels of difficulty eating/drinking solids and liquids. The study indicated that approximately two-thirds of participants reported difficulty swallowing liquids with 7/50 (14%) modifying liquid consistencies (n = 50). A slightly lower number reported difficulties with solids (31/50, 62%) with only 4/50 (8%) modifying solid consistencies.

The Dysphagia Severity Score [21,22] was used to assess participants' subjective perception of dysphagia severity in two studies [21,22]. Alfonsi et al [22] found that participants had a median score of two out of a possible two indicating that the majority of included participants reported severe swallowing difficulties (n = 9). A second included study also used the Dysphagia Severity Score [21]. Median scores were not available. However, participants reported a mean score of 1.4 indicating moderate-severe difficulties (n = 9).

The National Institutes of Health Speech Pathology Swallowing Questionnaire [38] was used in one study [20] (n = 27). This questionnaire addressed 20 possible swallowing difficulties. However, it does not provide an overall score. All participants had at least one complaint and the most common complaints were "coughing or choking", "excessive saliva," "difficulty swallowing," and "food falling out of the mouth".

Finally, a case report [23] used the Swallowing Disturbance Questionnaire [37] to assess patient reported swallowing difficulties. The participant scored 5/45 suggesting the participant's perceived level of swallowing difficulty was minimal (n = 1).

3.4.3. Other non-validated clinical swallowing assessments

There was variability in the description of CSE in included studies and inconsistent outcomes were reported meaning it was not possible to combine findings from different studies. Sulena et al [34] used the 3-ounce water swallow test [39] to identify possible dysphagia. Results indicated that 10/25 (40%) of the participants had water swallowing speed less than 10 ml/sec. Only one participant in this study was observed to cough during this screening tool.

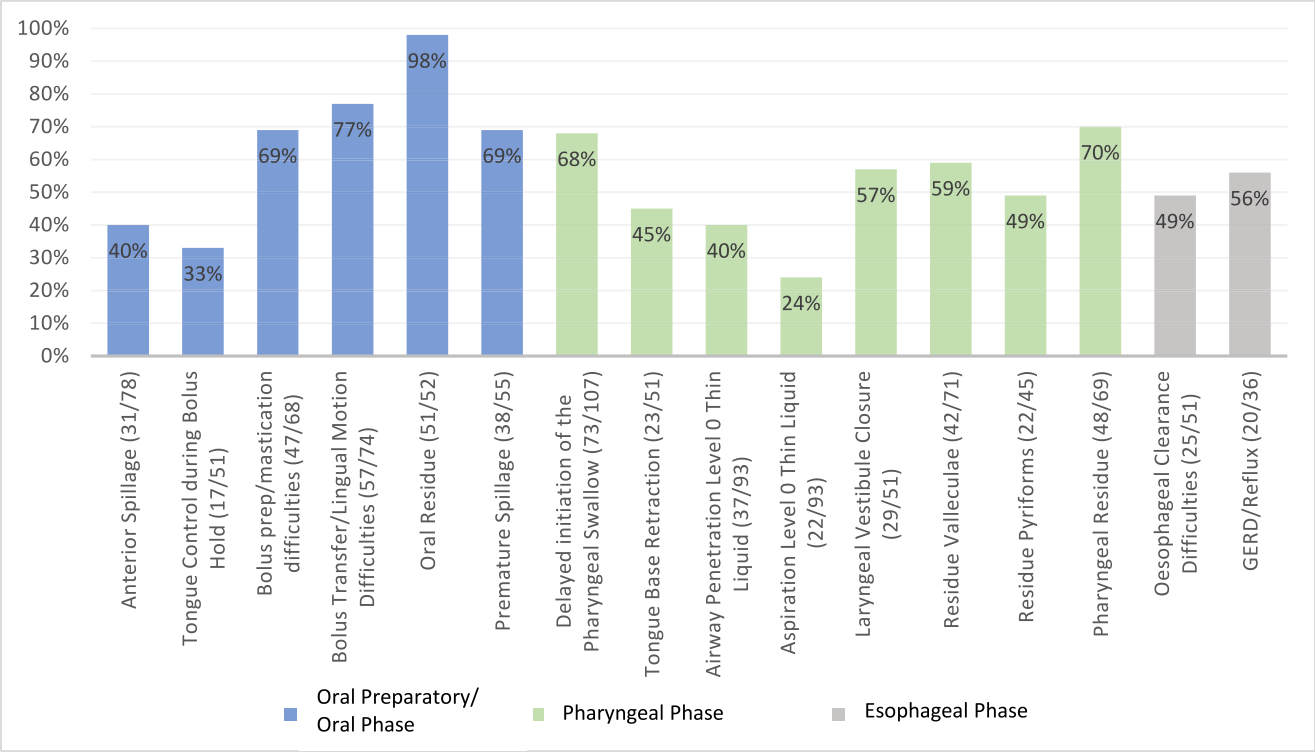

Fig. 2. Characteristics of dysphagia as described using instrumental assessment in included studies.

3.5. Differences in dysphagia presentation according to subtype of PSP

Of the 20 included studies, less than half (8/20) described subtypes of PSP. Of the 13 studies which used instrumental assessment, only four described characteristics according to subtype of PSP [15,16,27,28]. Of note, most participants in these studies had a diagnosis of PSP-RS (55/82, 67%) followed by PSP-P (15/82, 18%). As these studies used variable methods of assessment and outcome measures, it was not possible to combine findings.

Two studies using the PSPRS [36] provided scores for dysphagia items according to PSP subtypes [31,17]. The majority of participants in these studies had a diagnosis of PSP-RS (531/617, 86%), followed by PSP-P (45/617, 7%) and PSP-PGF (32/617, 5%).

For Question 13 on the PSPRS relating to dysphagia for liquids, combined scores from studies showed that participants with a diagnosis of PSP-RS presented with a median score of one out of a possible four [31,17] (mean = 1.09) (n = 531) indicating that most participants had to take "single sips, or fluid pools in mouth or pharynx, but no choking/coughing" was noted [36]. Similarly, the median score for Question 13 from the PSPRS for participants with a diagnosis of PSP-P was one (mean = 1.1) (n = 46) suggesting that participants with a diagnosis of PSP-RS and PSP-P present with similar characteristics of dysphagia relating to liquids. In contrast, participants with a diagnosis of PSP-PGF presented with lower scores for this question (median = 0; mean = 0.7) (n = 33).

For Question 3 on PSPRS relating to dysphagia for solids, median scores were available from one study [17]. Participants with a diagnosis of PSP-RS presented with a median score of 0/4 (mean = 0.62) (n = 465) [17] suggesting participants had no difficulties eating a full range of food textures. Similarly, participants with a diagnosis of PSP-P (n = 17) and PSP-PGF (n = 3) both had a median score of 0 (PSP-P mean = 0.4; PSP-PGF mean = 0.7) [17]. Results for other subtypes are not presented due to the under representation of these subtypes in included studies.

3.6. Impact of dysphagia on quality of life

While several studies used questionnaires or patient-reported interviews to identify the presence or describe the severity of dysphagia, only one study included a QOL assessment which incorporated dysphagia elements [29]. Piot and colleagues [29] used the PSP-Quality of Life Scale [40] which includes two questions relating to eating and swallowing. A breakdown of scores were not available for these questions. No studies used an assessment tool specifically evaluating the impact dysphagia has on QOL in people with PSP.

4. Discussion

This scoping review included 20 studies which described key methods for identifying and diagnosing dysphagia and outlined characteristics of dysphagia in people with PSP. This review found that a range of methods are being used to identify and diagnose dysphagia in PSP. The most commonly reported characteristics of dysphagia are oral preparatory and oral phase difficulties. Few studies included a breakdown of characteristics according to dysphagia subtype and only one study in this review evaluated the impact dysphagia has on QOL for people with PSP.

The majority of included studies in this scoping review used case control designs and were completed in Europe and North America highlighting the potential bias in research in PSP and dysphagia. For studies where severity of PSP was recorded, most participants had mild-moderate PSP highlighting a gap in research and may explain why most participants reported minimal swallowing difficulties for certain measures such as the PSPRS rating scale. These findings are in line with a recent study in dysphagia and IPD which highlighted the need to include underserved groups in research such as those with severe IPD to ensure generalizability of results [41]. This is also applicable for PSP where studies should include those with severe PSP and include a range of participants.

We found variable methods of assessment are used in identifying and diagnosing dysphagia in PSP. Assessment methods ranged from validated to non – validated subjective measures including instrumental assessments, patient-reported measures, clinical swallow evaluations and swallow screening tools. The evidence suggests the most-used instrumental assessments for dysphagia in this group are VFSS and FEES. The growing use of these tools will assist in more accurate and comprehensive assessment of dysphagia in PSP. However, the lack of standardized assessment procedures and protocols makes it difficult to compare findings between studies. It also means that reported symptoms of dysphagia will differ depending on the assessment type and outcomes used. This is reflected in the wide range of characteristics reported in the literature.

Despite this, there were a number of characteristics of dysphagia which appear to present more frequently in this cohort. Oral and pharyngeal residue, delayed initiation of the pharyngeal swallow and GERD/reflux are frequently reported dysphagia symptoms in the literature. Although further research is needed, given the early onset of dysphagia in this group, these characteristics could potentially act as indicators of PSP when patients are presenting with initial symptoms but have yet to receive a diagnosis [10].

The findings from this scoping review have implications for clinical practice. The results highlight the need for early and comprehensive dysphagia assessment including an instrumental assessment in patients with PSP. Given the frequency of oral preparatory and oral phase difficulties in people with PSP found in this review, dysphagia assessment should include an evaluation of this phase of swallowing for example by using VFSS. Robust and early dysphagia evaluation could potentially contribute to a timelier diagnosis and optimal management for patients. As a result of the rapid progression of dysphagia in this group, assessments at multiple timepoints are essential to track changes in swallow function which may be further indicators of a diagnosis of PSP [1].

Studies included in this scoping review did not consistently describe the subtypes of PSP. Of those studies which did describe PSP subtype, most participants had a diagnosis of PSP-RS. While it is possible that it highlights a lack of comprehensive assessment for this cohort, it is in line with evidence in PSP. Until recent years, most research has been based on PSP-RS and PSP-P [42]. The relatively recent introduction of the MDS criteria for the diagnosis of types of PSP [8] means that only studies since 2017 could have considered the subtypes outlined by Höglinger et al [8]. The lack of inclusion of subtypes other than PSP-RS means that it is difficult to tailor dysphagia care according to a specific subtype of PSP rather than providing identical care for all patients with PSP. There was some evidence suggesting that patients with PSP-RS and PSP-P presented with worse dysphagia symptoms than those with a diagnosis of PSP-PGF. However, the under-representation of subtypes other than PSP-RS means that these findings must be interpreted with caution. Future research should ensure subtypes of PSP and medical assessments such as neuroimaging results for each group are recorded in order to identify dysphagia symptoms that may be characteristic of particular subtypes.

Only one study described the impact dysphagia has on QOL in individuals with PSP. It is known that dysphagia can have a significant impact on a person's life in many other patient groups [43,44]. Future research should include this as a measure to ensure that a comprehensive and holistic perspective of a person's well-being is considered. Other consequences of dysphagia should also be considered including the impact dysphagia has on caregivers and alternative feeding.

5. Strengths and limitations

A strength of this review is that it sought to include all studies which described methods for identifying and diagnosing dysphagia in PSP, and studies which reported characteristics of dysphagia in PSP, with some

integration of unpublished data. While there were no limits placed on time of publication or language, as with all reviews some studies may have been missed despite our efforts to be as comprehensive as possible.

The heterogeneity of data reported in included studies led to challenges in collating the evidence. It was difficult to group characteristics as different assessment tools were used as well as variable outcome measures. As a result, this scoping review reported characteristics that were described for a minimum number of participants.

5.1. Conclusion

In conclusion, a range of assessment methods are used to identify and diagnose dysphagia in patients with PSP. A number of key dysphagia characteristics were evident in this group. The most frequently reported characteristics of dysphagia were oral preparatory and oral phase difficulties specifically oral residue and bolus transfer/lingual motion difficulties. Further research is needed to investigate if particular characteristics could be associated with certain subtypes. Future studies should also strive to include a measure of the impact dysphagia has on QOL.

CRediT authorship contribution statement

Éadaoin Flynn: Writing – review & editing, Writing – original draft, Visualization, Investigation, Formal analysis, Data curation, Conceptualization. **Julie Regan:** Writing – review & editing, Supervision, Investigation. **Julia Glinzer:** Writing – review & editing, Investigation, Formal analysis. **Sean O'Dowd:** Writing – review & editing, Supervision, Conceptualization. **Margaret Walshe:** Writing – review & editing, Supervision, Project administration, Methodology, Investigation, Conceptualization.

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Declaration of competing interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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Appendix A. Supplementary data

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References

[1] M. Roach, J.W. Chou, B. Lavin, J.R. Maclean, Challenges and opportunities in atypical parkinsonian syndromes: Call to action, Res. Rev. Parkinsonism 10 (2020) 1–6, https://doi.org/10.2147/JPRLS.S228108.

[2] E. Viscidi, I. Litvan, T. Dam, M. Juneja, L. Li, H. Krzywy, S. Eaton, S. Hall, J. Kupferman, G.U. Höglinger, Clinical features of patients with progressive supranuclear palsy in an US Insurance claims database, Front. Neurol. 12 (2021) 571800, https://doi.org/10.3389/fneur.2021.571800.

[3] J.C. Steele, J.C. Richardson, J. Olszewski, Progressive supranuclear palsy: A heterogeneous degeneration involving the brain stem, basal ganglia and cerebellum with vertical gaze and pseudobulbar palsy, nuchal dystonia and dementia, Semin. Neurol. 34 (2014) 129–150, https://doi.org/10.1055/s-0034-1377058.

[4] D.R. Williams, R. de Silva, D.C. Paviour, A. Pittman, H.C. Watt, L. Kilford, J. L. Holton, T. Revesz, A.J. Lees, Characteristics of two distinct clinical phenotypes in pathologically proven progressive supranuclear palsy: Richardson's syndrome and PSP-parkinsonism, Brain 128 (2005) 1247–1258, https://doi.org/10.1093/brain/awh488.

[5] U. Nath, Y. Ben-Shlomo, R. Thomson, H.R. Morris, N. Wood, A. Lees, D. Burn, The prevalence of progressive supranuclear palsy (Steele–Richardson–Olszewski syndrome) in the UK, Brain 124 (2001) 1438–1449, https://doi.org/10.1093/brain/124.7.1438.

[6] A. Schrag, Y. Ben-Shlomo, N.P. Quinn, Prevalence of progressive supranuclear palsy and multiple system atrophy: A cross-sectional study, Lancet 354 (1999) 1771–1775, https://doi.org/10.1016/s0140-6736(99)04137-9.

[7] I.T. Coyle-Gilchrist, K.M. Dick, K. Patterson, P.V. Rodríquez, E. Wehmann, A. Wilcox, C.J. Lansdall, K.E. Dawson, J. Wiggins, S. Mead, Prevalence, characteristics, and survival of frontotemporal Lobar degeneration syndromes, Neurol. 86 (2016) 1736–1743, https://doi.org/10.1212/WNL.0000000000002638.

[8] G.U. Höglinger, G. Respondek, M. Stamelou, C. Kurz, K.A. Josephs, A.E. Lang, B. Mollenhauer, U. Müller, C. Nilsson, J.L. Whitwell, T. Arzberger, E. Englund, E. Gelpi, A. Giese, D.J. Irwin, W.G. Meissner, A. Pantelyat, A. Rajput, J.C. van Swieten, I. Litvan, Clinical diagnosis of progressive supranuclear palsy: The Movement Disorder Society criteria, Mov. Disord. 32 (2017) 853–864, https://doi.org/10.1002/mds.26987.

[9] M.E. Finger, L.L. Madden, I.U. Haq, C.J. McLouth, M.S. Siddiqui, Analysis of the prevalence and onset of dysphonia and dysphagia symptoms in movement disorders at an academic medical center, J. Clin. Neurosci. 64 (2019) 111–115, https://doi.org/10.1016/j.jocn.2019.03.043.

[10] J. Müller, G.K. Wenning, M. Verny, A. McKee, K.R. Chaudhuri, K. Jellinger, W. Poewe, I. Litvan, Progression of dysarthria and dysphagia in postmortem-confirmed parkinsonian disorders, Arch. Neurol. 58 (2001) 259–264, https://doi.org/10.1001/archneur.58.2.259.

[11] D. Carneiro, M. das Gracas, W. de Sales Coriolano, L. Rodrigues Belo, A. Rocha de Marcos Rabelo, A. Guescel Asano, O. Gomes Lins, Quality of life related to swallowing in Parkinson's disease, Dysphagia. 29 (2014) 578–582, https://doi.org/10.1007/s00455-014-9548-3.

[12] C. dell'Aquila, S. Zoccolella, V. Cardinali, M. de Mari, G. Iliceto, B. Tartaglione, P. Lamberti, G. Logroscino, Predictors of survival in a series of clinically diagnosed progressive supranuclear palsy patients, Parkinsonism Relat. Disord. 19 (2013) 980–985, https://doi.org/10.1016/j.parkreldis.2013.06.014.

[13] A.C. Tricco, E. Lillie, W. Zarin, K.K. O'Brien, H. Colquhoun, D. Levac, D. Moher, M. Peters, T. Horsley, L. Weeks, S. Hempel, E.A. Akl, C. Chang, J. McGowan, L. Stewart, L. Hartling, A. Aldcroft, M.G. Wilson, C. Garritty, S.E. Straus, PRISMA Extension for Scoping Reviews (PRISMAScR): Checklist and Explanation, Ann. Intern. Med. 169 (2018) 467–473, https://doi.org/10.7326/M18-0850.

[14] E. Flynn, J. Regan, J. Radtke, S. O'Dowd, M. Walshe, Dysphagia in progressive supranuclear palsy: A scoping review protocol, Adv. Commun. Swallowing 25 (2022) 109–113, https://doi.org/10.3233/ACS-220007.

[15] J.C. Borders, J.S. Sevitz, J.A. Curtis, N. Vanegas-Arroyave, M.S. Troche, Quantifying impairments in swallowing safety and efficiency in progressive supranuclear palsy and Parkinson's disease, Dysphagia 38 (2023) 1342–1352, https://doi.org/10.1007/s00455-023-10560-7.

[16] H.M. Clark, J.A.G. Stierwalt, H. Tosakulwong Botha, F. Ali, J.L. Whitwell, K. A. Josephs, Dysphagia in progressive supranuclear palsy, Dysphagia 35 (2020) 667–676, https://doi.org/10.1007/s00455-019-10073-2.

[17] L.I. Golbe, P. Ohman-Strickland, E.B. Beisser, F.T. Elghoul, A Convenient prognostic tool and staging system for progressive supranuclear palsy, Mov. Disord. Clin. Pract. 7 (2020) 664–671, https://doi.org/10.1002/mdc3.13010.

[18] B.T. Johnston, J.A. Castell, S. Stumacher, A. Colcher, R.M. Gideon, Q. Li, D. O. Castell, Comparison of swallowing function in Parkinson's disease and progressive supranuclear palsy, Mov. Disord. 12 (1997) 322–327, https://doi.org/10.1002/mds.870120310.

[19] N.A. Leopold, M.C. Kagel, Dysphagia in progressive supranuclear palsy: radiologic features, Dysphagia 12 (1997) 140–143, https://doi.org/10.1007/PL00009528.

[20] I. Litvan, N. Sastry, B.C. Sonies, Characterizing swallowing abnormalities in progressive supranuclear palsy, Neurol. 48 (1997) 1654–1662, https://doi.org/10.1212/wnl.48.6.1654.

[21] E. Alfonsi, M. Versino, I.M. Merlo, C. Pacchetti, E. Martignoni, G. Bertino, A. Moglia, C. Tassorelli, G. Nappi, Electrophysiologic patterns of oral-pharyngeal swallowing in parkinsonian syndromes, Neurol. 68 (2007) 583–589, https://doi.org/10.1212/01.wnl.0000254478.46278.67.

[22] E. Alfonsi, I.M. Merlo, M. Ponzio, C. Montomoli, C. Tassorelli, C. Biancardi, A. Lozza, E. Martignoni, An electrophysiological approach to the diagnosis of neurogenic dysphagia: implications for botulinum toxin treatment, J. Neurol. Neurosurg. Psychiatry 81 (2010) 54–60.

[23] N. Beschin, C. Reverberi, S. Della Sala, Anosognosia for chronic dysphagia, Cortex: A J. Devoted to the Study of the Nerv, Syst. Behav. 109 (2018) 355–357, https://doi.org/10.1016/j.cortex.2018.08.023.

[24] G. Cosentino, C. Tassorelli, P. Prunetti, M. Todisco, R. De Icco, M. Avenali, B. Minafra, R. Zangaglia, F. Valentino, C. Pacchetti, G. Bertino, S. Mauramati, M. Fresia, E. Alfonsi, Reproducibility and reaction time of swallowing as markers of

dysphagia in parkinsonian syndromes, Clin. Neurophysiol. 131 (2020) 2200–2208, https://doi.org/10.1016/j.clinph.2020.06.018.
[25] M. Picillo, M.F. Tepedino, M.C. Russillo, F. Abate, M. Savastano, A. De Simone, R. Erro, M.T. Pellecchia, P. Barone, Energy expenditure, body composition and dietary habits in progressive supranuclear palsy, J. Neurol. May 269 (2022) 2610–2618, https://doi.org/10.1007/s00415-021-10846-6.
[26] S. Varanese, P. Di Ruscio, L. Ben M' Barek, A. Thomas, M. Onofrj, Responsiveness of dysphagia to acute L-Dopa challenge in progressive supranuclear palsy, J. Neurol. 261 (2014) 441-2, https://doi.org/10.1007/s00415-013-7232-4.
[27] I. Claus, J. Suttrup, P. Muhle, S. Suntrup-Krueger, M.L. Siemer, F. Lenze, R. Dziewas, T. Warnecke, Subtle esophageal motility alterations in parkinsonian syndromes: Synucleinopathies vs, Tauopathies, Mov. Disord. Clin. Pract. 5 (2018) 406–412, https://doi.org/10.1002/mdc3.12616.
[28] W. Maetzler, T.W. Rattay, M.A. Hobert, M. Synofzik, A. Bader, D. Berg, E. Schaeffer, N. Rommel, D. Devos, B.R. Bloem, B. Bender, Freezing of swallowing, Mov. Disord. Clin. Pract. 3 (2016) 490–493, https://doi.org/10.1002/mdc3.12314.
[29] I. Piot, K. Schweyer, G. Respondek, M. Stamelou, DescribePSP study group, ProPSP study group, MDS-endorsed PSP study group, P. Sckopke, T. Schenk, C.G. Goetz, G. T. Stebbins, G.U. Höglinger, The Progressive Supranuclear Palsy Clinical Deficits Scale, Mov. Disord. 35 (2020) 650-661, https://doi.org/10.1002/mds.27964.
[30] T. Warnecke, S. Oelenberg, I. Teismann, C. Hamacher, H. Lohmann, E. B. Ringelstein, R. Dziewas, Endoscopic characteristics and levodopa responsiveness of swallowing function in progressive supranuclear palsy, Mov. Disord. 25 (2010) 1239–1245, https://doi.org/10.1002/mds.23060.
[31] J.H. Choi, H. Kim, J.H. Shin, J.Y. Lee, H.J. Kim, J.M. Kim, B. Jeon, Eye movements and association with regional brain atrophy in clinical subtypes of progressive supranuclear palsy, J. Neurol. 268 (2021) 967–977, https://doi.org/10.1007/s00415-020-10230-w.
[32] I.J. Han, H.G. Kwon, W.W. Lee, R.G. Yoon, H. Choi, H.J. Kim, Diffusion tensor tractography of the corticobulbar tract in a dysphagic patient with progressive supranuclear palsy: A case report, Medicine 102 (2023) 32898–e32898, https://doi.org/10.1097/MD.0000000000032898.
[33] E. Kaphan, J.F. Pellissier, M. Rey, D. Robert, M. Auphan, A. Ali Chérif, Esophageal achalasia, sleep disorders and chorea in a tauopathy without ophthalmoplegia, parkinsonian syndrome, nor dementia (progressive supranuclear palsy?): Clinicopathological study, Rev. Neurol. (paris). 164 (2008) 377–383, https://doi.org/10.1016/j.neurol.2007.09.007.
[34] S. Sulena, D. Gupta, A.K. Sharma, B. Singh, Clinical profile of dysphagia in patients with Parkinson's disease, progressive supranuclear palsy and multiple system atrophy, J. Assoc. Physicians India 65 (2017) 32–37.
[35] M.M. Hoehn, M.D. Yahr, Parkinsonism: Onset, progression and mortality, Neurol. 17 (1967) 427–442, https://doi.org/10.1212/wnl.17.5.427.
[36] L.I. Golbe, P.A. Ohman-Strickland, A clinical rating scale for progressive supranuclear palsy, Brain 130 (2007) 1552–1565, https://doi.org/10.1093/brain/awm032.
[37] J.T. Cohen, Y. Manor, Swallowing disturbance questionnaire for detecting dysphagia, Laryngoscope 121 (2011) 1383–1387, https://doi.org/10.1002/lary.21839.
[38] B. Sonies, L. Parent, K. Monish, B. Baum, Durational aspects of the oral-pharyngeal phase of swallow in normal adults, Dysphagia 3 (1988) 1–10, https://doi.org/10.1007/BF02406274.
[39] D.M. Suiter, S.B. Leder, Clinical utility of the 3-ounce water swallow test, Dysphagia 23 (2008) 244–250, https://doi.org/10.1007/s00455-007-9127-y.
[40] A. Schrag, C. Selai, N. Quinn, A. Lees, I. Litvan, A. Lang, Y. Poon, J. Bower, D. Burn, J. Hobart, Measuring quality of life in PSP: the PSP-QoL, Neurol. 67 (2006) 39–44, https://doi.org/10.1212/01.wnl.0000223826.84080.97.
[41] J. Hirschwald, L. Finnegan, J. Hofacker, M. Walshe, Underserved groups in dysphagia intervention trials in Parkinson's disease: A scoping review, Ageing Res. Rev. 93 (2024), https://doi.org/10.1016/j.arr.2023.102150.
[42] P. Krzosek, N. Madetko, A. Migda, B. Migda, D. Jagus, P. Alster, Differential diagnosis of rare subtypes of progressive supranuclear palsy and PSP-like syndromes—infrequent manifestations of the most common form of atypical parkinsonism, Front. Aging Neurosci. 14 (2022) 804385, https://doi.org/10.3389/fnagi.2022.804385.
[43] R. Smith, L. Bryant, B. Hemsley, The true cost of dysphagia on quality of life: The views of adults with swallowing disability, Int. J. Lang. Commun. Disord. 58 (2023) 451–466, https://doi.org/10.1111/1460-6984.12804.
[44] L.P. Leow, M.L. Huckabee, T. Anderson, L. Beckert, 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 25 (2010) 216–220, https://doi.org/10.1007/s00455-009-9245-9.