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Protocol

Dysphagia in progressive supranuclear palsy: A scoping review protocol

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Abstract.
BACKGROUND: 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.
OBJECTIVE: 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.
METHODS: The proposed scoping review will be conducted in accordance with the JBI methodology for scoping reviews. The Preferred Reporting Items for Systematic Reviews and Meta Analysis extension for scoping reviews (PRISMA-ScR) will be used to guide the reporting of the review (Tricco et al., 2018). Articles completed at any time, which include participants with dysphagia and a clinical diagnosis of PSP will be included. Studies involving participants who have a co-morbidity/morbidities which could cause dysphagia and secondary research will be excluded. Relevant electronic databases, trial registries and grey literature without any date or language restrictions will be searched. Two independent reviewers will assess articles for eligibility and will extract relevant data.
CONCLUSIONS: This scoping review will provide important evidence on dysphagia and PSP. It will describe the principal methods used for identifying and diagnosing dysphagia in this population. The results will guide future research in dysphagia and PSP.

Keywords: Deglutition, PSP, parkinsonism, richardson syndrome, scoping review

1. Introduction

Atypical Parkinsonian syndromes (APS) encompass a collective of rare neurodegenerative diseases including progressive supranuclear palsy (PSP), multiple system atrophy (MSA), corticobasal syndrome (CBS), and dementia with Lewy Bodies (DLB). APS are characterised by rapid disease progression and decreased life expectancy (Roach et al., 2020). APS can be particularly debilitating and are not yet well understood. One of the most common APS is PSP.

PSP is typically characterised by postural instability, supranuclear vertical gaze palsy leading to impaired vision, dementia, dysarthria, and other features including swallowing disorders (dysphagia) (Viscidi et al., 2021; Steele, Richardson & Olszewski, 2014). Two subtypes of PSP have primarily been

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described in the literature, Richardson syndrome (PSP-RS) and PSP-parkinsonism (PSP-P) (Williams et al., 2005). PSP-RS is the most common type of PSP and is characterised 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 (Viscidi et al., 2021). In contrast, PSP-P is characterised by asymmetric onset, tremor, and moderate 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 presentation of PSP is estimated to be up to 6.4/100,000 (Nath et al., 2001; Schrag, Ben-Shlomo & Quinn, 1999). However, more recent research suggests that this figure may be higher, with average age of onset in the mid-60s and disease duration of approximately 6 years (Coyle-Gilchrist et al., 2016).

In recent years, multiple variants of PSP have been described by The Movement Disorder Society (MDS) based on initial clinical presentations (Hoglinger et al., 2017). The MDS outlined new criteria aimed at optimising early, sensitive and specific clinical diagnosis of PSP based on current evidence (Höglinger et al., 2017). Initial clinical predominance types include PSP-RS (Richardson syndrome), PSP-OM (ocular motor dysfunction), PSP-PI (postural instability), PSP-P (parkinsonism resembling IPD), PSP-F (frontal lobe cognitive or behavioural presentations), PSP-PGF (progressive gait freezing), PSP-CBS (corticobasal syndrome) and PSP-SL (speech/language disorders) (Höglinger et al., 2017). As the disease progresses, subtypes may evolve into PSP-RS.

Dysphagia is common in patients with PSP yet it is underdiagnosed and methods of assessing dysphagia vary (Finger et al., 2019). PSP is associated with early onset of dysphagia when compared with IPD (Clark et al., 2020). Dysphagia can result in clinical complications, including malnutrition, dehydration, and aspiration pneumonia, affecting quality of life and eventually increasing the mortality rate in these patients. Aspiration pneumonia secondary to dysphagia is a major risk in advanced PSP and is the principal cause of death (dell'Aquila et al., 2013).

To date, many studies have focused on IPD and dysphagia however, research investigating dysphagia in PSP is scant. 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 potential consequences of dysphagia in this population 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 quality of life in this population will inform future research which may aid in earlier diagnosis of PSP. An improved understanding of the nature, diagnostic methods and impact dysphagia has on individuals with all subtypes of PSP can ultimately optimise dysphagia management and hence reduce adverse clinical and quality of life outcomes.

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

1.1. Review questions

- 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 are the key methods for identifying and diagnosing dysphagia in this population?
- What impact does dysphagia have on quality of life in people with PSP?

2. Methods

The proposed scoping review will be conducted in accordance with the JBI methodology for scoping reviews (Tricco et al. 2018). The Preferred Reporting Items for Systematic Reviews and Meta Analysis extension for scoping reviews (PRISMA-ScR) will be used to guide the reporting of the review (Tricco et al., 2018). A preliminary search of MEDLINE, PROSPERO, the Cochrane Database of Systematic Reviews and JBI Evidence Synthesis was conducted and no current or underway systematic reviews or scoping reviews on the topic were identified.

3. Eligibility criteria

3.1. Participants

Table 1 contains the eligibility criteria for the scoping review. Studies which include participants with

Table 1
Inclusion and Exclusion Criteria

Inclusion Criteria	Exclusion Criteria
– Studies which include participants with a clinical diagnosis of PSP established by a neurologic exam and dysphagia of any type or severity will be included.	Studies where participants have a co-morbidity/co-morbidities which could also cause dysphagia.
– RCTs, non-RCTs, before and after studies and interrupted time-series studies. – Prospective and retrospective cohort studies, case-control studies and analytical cross-sectional studies. – Case series, individual case reports and descriptive cross-sectional studies. – Qualitative studies. – Systematic reviews. – Grey literature including conference posters and presentations.	Editorials, expert opinions and secondary research e.g. literature reviews.
– Studies in any language.	
– Studies completed since inception to March 2023.	
– Studies completed in any setting.	

a clinical diagnosis of PSP established by a neurologic exam and dysphagia of any type or severity will be included. Studies will be excluded if participants have a co-morbidity or comorbidities which could independently cause dysphagia.

3.2. Types of sources

Research based in any country and in any setting will be included in this review. Studies published since inception to March 2023 and, in any language, will be included. This scoping review will consider both experimental and quasi-experimental study designs including randomised controlled trials (RCTs), non-randomised 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 will be considered for inclusion. This review will also consider descriptive observational study designs including case series, individual case reports and descriptive cross-sectional studies for inclusion.

Qualitative studies that focus on qualitative data including, but not limited to, designs such as phenomenology, grounded theory, ethnography, qualitative description and action research will also be included. In addition, systematic reviews that meet the inclusion criteria will also be considered. Grey literature including conference posters and presentations will also be included if sufficient detail is available to address the research questions. Editorials, expert opinions and secondary research for example, literature reviews will be excluded.

3.3. Search strategy

The search strategy will aim to identify both published and unpublished studies. An initial 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 for relevant databases and information sources with the assistance of a subject librarian (See Appendix I). The search strategy, including all identified keywords and index terms, will be adapted for each included database and/or information source. The reference lists of all included sources of evidence will be screened for additional studies. The databases to be searched include PubMed, EMBASE, CINAHL, Web of Science Core Collection and PsycINFO. Sources of unpublished studies/grey literature to be searched include ProQuest Dissertation and Theses Global.

3.4. Selection of studies

Following the search, all identified citations will be collated and uploaded into an online platform (www.covidence.org) and duplicates will be removed. Following a pilot test, titles and abstracts will then be screened by two independent reviewers (EF, JR) for assessment against the inclusion criteria for the review. Potentially relevant sources will be retrieved in full, and their citation details imported into Covidence (www.covidence.org). The full text of selected citations will be assessed in detail against the inclusion criteria by the two independent reviewers. Reasons for exclusion of sources of evidence at

full text that do not meet the inclusion criteria will be recorded and reported in the scoping review. Any disagreements that arise between the reviewers at each stage of the selection process will be resolved through discussion, or with additional reviewers (MW, JaR). The results of the search and the study inclusion process will be reported in full in the final scoping review and presented in a PRISMA-ScR flow diagram (Tricco et al., 2018).

3.5. Data extraction

Data will be extracted from papers included in the scoping review by two independent reviewers using a data extraction tool developed by the reviewers (See Appendix II) and an online platform (www.covidence.org). The data extracted will include specific details about the participants, concept, context, study methods and key findings relevant to the review questions. The data extraction form will be piloted independently on three included studies by EF and JR. Following this, the data extraction form will be modified and revised as necessary during the pilot process. Any changes will be described in the scoping review. Any disagreements that arise between the reviewers will be resolved through discussion, or with additional reviewers (JaR, MW). If required, authors of papers will be contacted to request missing or additional data for studies completed within the past five years.

3.6. Data analysis and presentation

Data from each article will be presented in written and visual format. Depending on the quantity of papers included, where possible, data relating to dysphagia will be categorised into oral preparatory/oral stage difficulties, pharyngeal stage difficulties and/or oesophageal stage difficulties.

Identification of methods for diagnosing dysphagia in this population will be discussed including potential benefits and limitations. Methods of dysphagia assessment will be categorised into swallow screening, clinical (e.g. clinical swallow evaluation) and instrumental (e.g. fibreoptic endoscopic evaluation of swallowing, videofluoroscopy, high resolution pharyngeal manometry) assessment methods. Patient reported outcome measures will be used to describe the impact dysphagia has on quality of life in people with PSP. Differences in dysphagia presentation according to the subtype of PSP will be analysed.

4. Discussion

This scoping review will increase our knowledge of dysphagia in PSP. It aims to identify differences in dysphagia presentation according to the subtype of PSP, outline diagnostic methods and the impact dysphagia has on quality of life in this population. Variables such as cognition and clinical complications associated with dysphagia in PSP are outside of the scope of this paper. However, these factors may be examined in future research. The results from this review will have implications for research and clinical practice. A better understanding of dysphagia in all subtypes of PSP can ultimately optimise dysphagia management, facilitate earlier medical diagnosis and improve clinical outcomes and quality of life. Furthermore, the results will identify gaps in the literature and guide future research.

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Conflict of interest

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Supplementary material

The Appendix I and II is available in the electronic version of this article: https://dx.doi.org/10.3233/ACS-220007.

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