IV SESSIONE: PROBLEMI DIAGNOSTICI E TERAPEUTICI NELLE DISFAGIE NEUROGENE

La gestione riabilitativa delle disfagia nelle malattie degenerative del Sistema Nervoso Centrale

Dr.ssa Micol Avenali
University of Pavia, Dept. Brain and Behavioral Science, Pavia - IT
IRCCS Mondino Foundation, Pavia - IT
Parkinson’s Disease

Atypical parkinsonism (MSA)

Huntington’s disease

MANIFESTAZIONE CLINICA

APPROCCIO TERAPEUTICO

VALUTAZIONE STRUMENTALE
Dysphagia in Neurodegenerative Disease (ND) is still too frequently underdiagnosed.

Guidelines for screening, diagnosis and management of dysphagia in ND are lacking.

**International Consensus Conference on PD**
Systematic revision of the literature published from January 1990 to February 2021

**International Consensus Conference on MSA**
Systematic revision of the literature published from January 1990 to May 2020

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**A multinational consensus on dysphagia in Parkinson's disease: screening, diagnosis and prognostic value**

Giuseppe Cosentino1,2, Micol Avenali1,3, Antonio Schindler4, Nicole Pizzorni5, Cristina Montomoli6, Giovanni Abbruzzeze6, Angelo Antonini1, Filippo Barbiera8, Marco Benazzo9, Eduardo Elias Benarroch10, Giulia Bertino3, Emanuele Cerdia11, Pere Clavé12,13, Pietro Cortelli14,15, Roberto Eleopra16, Chiara Ferrari16, Shaheen Hamdy17, Maggie-Lee Huckabee18, Leonardo Lopiano19, Rosario Marchese-Ragona20, Stefano Masiero21, Emilia Michou22, Antonio Occhini23, Claudio Pacchetti23, Ronald F. Pfeffer24, Domenico A. Restivo25, Mariangela Rondanelli26, Giovanni Ruoppolo27, Giorgio Sandrini1, Anthony H. V. Schapira28, Fabrizio Stocchi29, Eduardo Tolosa30, Francesca Valentino31, Mauro Zamboni31, Roberta Zangaglia23, Mario Zappia22, Cristina Tassorelli1,3, Enrico Alfonsi2

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**Review article**

Dysphagia in multiple system atrophy consensus statement on diagnosis, prognosis and treatment

SCREENING
When should dysphagia be assessed?

DIAGNOSIS
How should dysphagia be assessed?

TREATMENT
What are the treatment options for dysphagia?

PROGNOSIS
Does dysphagia influence the prognosis?
DYSPHAGIA IN PARKINSON’S DISEASE

- Dysphagia is a common symptom in PD
- The prevalence of dysphagia in PD is estimated between 40 and 80%
- It may involve oral, pharyngeal or esophageal phases
- It may be present in every stage of the disease
- It worsens with disease progression and may vary with motor fluctuations
- It can negatively affect the quality of life
- It can result in severe complications and hospitalization
A multinational consensus on dysphagia in Parkinson's disease: screening, diagnosis and prognostic value

Giuseppe Cosentino1,2, Micol Avenali1,2, * Antonio Schindler4, Nicole Pizzorni4, Cristina Montomoli2, Giovanni Abbuzzese5, Angelo Antonini1, Filippo Barbieri6, Marco Benazzi7, Eduardo Elias Benarroch8, Giulia Bertino9, Emanuele Cereda1, Piero Clavel10,11, Pietro Cortelli11,12, Roberto Eleopra12, Chiara Ferrari11, Shaheen Hamdy17, Maggie-Lee Huckabees18, Leonardo Lopiano19, Rosario Marchese Ragona20, Stefano Masiero21, Emilia Michou22, Antonio Occhini14, Claudio Pacchetti23, Ronald F. Pfeiffer24, Domenico A. Restivo25, Mariangela Rondanelli26, Giovanni Ruoppolo27, Giorgio Sandrini1, Anthony H. V. Schapira28, Fabrizio Stocchi29, Eduardo Tolosa30, Francesca Valentino23, Mauro Zamboni21, Roberta Zangaglia31, Mario Zappa22, Cristina Tassorelli12, Enrico Alfonsi2

Table 1 Descriptive features of the 117 eligible studies included in the evaluation of screening, diagnosis, QoL and prognosis

<table>
<thead>
<tr>
<th>Topic domain</th>
<th>All studies</th>
<th>Number of studies</th>
<th>Cross-sectional/prospective studies</th>
<th>Case–control/retrospective study</th>
<th>Class of evidence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Screening and diagnosis of dysphagia in PD</td>
<td>82</td>
<td>Number of studies</td>
<td>58</td>
<td>24</td>
<td>4 Class I</td>
</tr>
<tr>
<td></td>
<td></td>
<td>N</td>
<td>N</td>
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<td>N</td>
</tr>
<tr>
<td></td>
<td>&lt; 10 pts = 1</td>
<td>10–19 pts = 16 studies</td>
<td>20–50 pts = 28 studies</td>
<td>&gt; 50 pts = 37 studies</td>
<td>4 Class II</td>
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<tr>
<td></td>
<td>(n = 61)</td>
<td>(n = 24)</td>
<td>18 Class III</td>
<td>56 Class IV</td>
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</tbody>
</table>
HOW TO ASSESS DYSPHAGIA IN PD?

No standardized protocols have been developed for the screening and diagnosis of dysphagia in PD

Need for an early identification of swallowing abnormalities in PD

QUESTIONS ON SCREENING

- When is it indicated to screen for dysphagia in patients with PD?
- When should dysphagia be suspected in patients with PD?
- What clinical tools should be used to screen for dysphagia in patients with PD?

QUESTIONS ON DIAGNOSIS

- What clinical tools should be used to detect the presence of dysphagia?
- What instrumental investigations should be used to detect the presence of dysphagia?
- How should severity of dysphagia be assessed?
**When is it indicated to screen for dysphagia in patients with PD?**

Search for symptoms or signs that are suspicious for the presence of dysphagia:

- increased duration of meals
- difficulty in tablet swallowing
- sensation of food sticking in the throat after swallowing
- coughing and choking during ingestion of food and liquids
- gurgling voice
- weight loss
- low body mass index
- recurrent chest infections
- drooling

**When should dysphagia be suspected in patients with PD?**

- The risk of dysphagia increases with the number of symptoms or signs observed, age, and disease progression.

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**A multinational consensus on dysphagia in Parkinson’s disease: screening, diagnosis and prognostic value**

Giuseppe Cosentino1,3, Michel Avena1,70, Antonio Schindler4, Nicola Pizzorni5, Cristina Montomoli8, Giovanni Abbazzese9, Angelo Antonini10, Filippo Barbieri11, Marco Benazzo12, Eduardo Elias Benarroch13, Giulia Bertino14, Emanuele Cereda15, Pierre Clavel12,15, Pietro Cortell11,15, Roberto Eleopa16, Chiara Ferrari17, Shahen Hamdy18, Maggie-Lee Huckabee19, Leonardo Lopiano11, Rosario Marchese Ragona20, Stefano Massiro11, Emilia Michou11, Antonio Ochini1, Claudio Pacchetti21, Ronald F. Pfeffer22, Domenico A. Restivo23, Mariangela Rondanelli24, Giovanni Ruoppolo25, Giorgio Sandrini1, Anthony H. V. Schapira26, Fabrizio Stocchi27, Eduardo Tolosa28, Francesca Valentino29, Mauro Zamboni30, Roberta Zangaglia31, Mario Zappia32, Cristiana Tassorelli14, Enrico Alfonsi7
### Scales and questionnaires used to assess Dysphagia in PD

- **Swallowing disturbance questionnaire (SDQ)**
  - Self-reported questionnaire containing 15 items on swallowing
  - Detects early dysphagia in PD
  - 80.5% sensitivity and 81.3 % specificity

- **Munich Dysphagia test-Parkinson’s disease (MDT-PD)**
  - Self-reported questionnaire (26 items)
  - Detects initial oropharyngeal symptoms and the risk of laryngeal penetration and/or aspiration in PD

- **Swallowing Clinical Assessment Score in Parkinson’s disease (SCAS-PD)**
  - Quantitative clinical scale (12 items)
  - Detects alterations in the oral and pharyngeal phases of swallowing
  - Good sensitivity/ specificity and concordance with the VFSS

- **Radboud Oral Motor Inventory for Parkinson’s disease (ROMP)**
  - Questionnaire that assesses three speech, swallowing, and saliva control
  - Valid tool to identify swallowing difficulties in PD
  - Contains a limited number of items

- **Handheld Cough Testing (HCT)**
  - Tool for cough and dysphagia assessment in PD
  - Identifies differences in cough airflow during reflex and voluntary cough tasks
  - High sensitivity and specificity
From DYMUS to DYPARK: Validation of a Screening Questionnaire for Dysphagia in Parkinson’s Disease

Carlotta Dagna1,2, Micol Avenali1,2, Roberto De Icco1,2, Marialuisa Gandolfi3, Claudio Solaro4, Domenico Restivo4, Michelangelo Bartolo4, Francesca Meneghelli7, Giorgio Sandrini1,2, Cristina Tassorelli1,2

DYPARK SIRN Group

Observational multicentric study involving 103 PD

<table>
<thead>
<tr>
<th>Variable</th>
<th>Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>N. Subjects</td>
<td>103</td>
</tr>
<tr>
<td>Sex (M/F)</td>
<td>56/47</td>
</tr>
<tr>
<td>Age (years), mean ± SD</td>
<td>68.0± 2.8</td>
</tr>
<tr>
<td>Disease duration (years), mean ± SD</td>
<td>11.5± 4.9</td>
</tr>
</tbody>
</table>

DYMUS Questionnaire

1. Do you have difficulty swallowing solid food (such as meat, bread)? Yes □ No □
2. Do you have difficulty swallowing liquids (such as water, milk)? Yes □ No □
3. Do you have a globus sensation (the feeling of a lump) in your throat when swallowing? Yes □ No □
4. Does food stick in your throat? Yes □ No □
5. Do you cough or have a choking sensation after ingesting solid food? Yes □ No □
6. Do you cough or have a choking sensation after ingesting liquids? Yes □ No □
7. Do you need to swallow several times before solid food “goes down” completely? Yes □ No □
8. Do you need to cut food into small pieces be able to swallow it? Yes □ No □
9. Do you need to take many sips in order to drink? Yes □ No □
10. Have you lost weight? Yes □ No □
ROC curves for the ability of the DYMUS Scale to identify PD patients with an increased risk of dysphagia. We identified the DYMUS score of 6 as the cut-off for identifying patients at higher risk of dysphagia with a sensibility of 0.7 and a specificity of 0.84.

**DYMUS → DYPARK** proved to be a reliable screening tool to detect dysphagia in PD
What clinical tools should be used to detect the presence of dysphagia?

- **Clinical swallowing examination**
  - Examination of cranial nerves
  - Evaluation of dry swallows on-command and/or reflexive cough testing
  - Evaluation of swallowing of various food and liquid consistencies
  - Detection of possible signs or symptoms of reduced swallowing efficiency

- **Assessment of cognition and speech**
  - *Swallowing examination should be performed during an ON phase*
  - *Clinical examination should not be performed during in the presence of cervical-cranial dyskinesias*
  - *Patients with DBS implants should be tested in an ON medication/ ON stim*
  - *Evaluate possible interactions between DBS and swallowing*
What clinical tools should be used to detect the presence of dysphagia?

- Meal Observation
  - Water Swallow Test (WST)
  - Voluntary and/or reflex cough test
- Assessing a higher number of swallowing acts
- Evaluate meal duration

A multinational consensus on dysphagia in Parkinson’s disease: screening, diagnosis and prognostic value

What instrumental investigations should be used to detect the presence of dysphagia?

**Videofluoroscopic swallow study (VFSS)**
- Directly reveals penetration/aspiration
- Provides information when the oral and/or esophageal phase is impaired
- Less easier to perform
- Requires the use of radiation
- Can not be repeated at short time intervals

**Fiberoptic endoscopic evaluation of swallowing (FEES)**
- Assessment of the pharyngeal phase
- Detection of penetration/aspiration phenomena
- Easier to perform (i.e., patient’s bedside)
- Allows to test swallowing of real food
- Does not require the use of radiation
- It can be repeated at short time intervals
- Can be used in follow-ups

Enable detection of aspiration, penetration, and residue with high sensitivity and specificity
The electrophysiological evaluation of oropharyngeal swallowing might provide further insights into the pathophysiological basis of dysphagia in PD and give useful clues for treatment.

Gastroenterological investigations:

- Pharyngo-esophageal manometry -HRM
- Upper gastrointestinal endoscopy
- Barium swallow
- Acid/reflux-related test

Should always be carried out in the presence of esophageal symptoms.
How should severity of dysphagia be assessed?

No validated scales specific for PD to rate dysphagia severity. Swallowing severity should be carried out to guide the best treatment strategy and for prognostic assessment.

- **Penetration-Aspiration Scale (PAS)**
  - Based on imaging data
  - Most adopted

- **Dysphagia Outcome and Severity Scale (DOSS)**
  - Based on clinical and instrumental parameters

- **Functional Oral Intake Scale (FOIS)**
  - Based on clinical assessment
Clinical or instrumental evidence of impairment of:

❖ swallowing safety

AND/OR

❖ Swallowing efficiency

TREATMENT SHOULD BE STARTED

Optimization of PD treatments

Dietary modifications

Swallowing therapy

Botulinum Toxin injection

Neuromodulation

BOLUS ADAPTATION
- volume
- viscosity
- temperature

FACILITATING TECHNIQUES
- lingual muscles
- velo-pharyngeal muscles
- cervical muscles

STRENGTHENING EXERCISES
- lingual muscles
- velo-pharyngeal muscles
- cervical muscles
QUALITY OF LIFE and PROGNOSIS

STATEMENTS ON QoL
- Dysphagia affects the QoL of PD patients
- Dysphagia severity seems to correlate with poorer QoL
- The three main domains of QoL affected by dysphagia in PD patients are:
  - loss of the social aspect of eating;
  - loss of personal autonomy;
  - difficulties in taking oral therapy.

STATEMENTS ON PROGNOSIS
- The presence of dysphagia negatively influences the prognosis of PD patients
- Aspiration in the lungs is strongly correlated to a higher risk of choking and aspiration pneumonia.
- Poor oral care, load of comorbidities and cognitive impairment are possibly associated to a worse prognosis in dysphagic PD patients.
**Atypical parkinsonism (MSA)**

**Statements on dysphagia treatment**

Class IV level studies and expert adaptation of knowledge from the literature on dysphagia

- Dysphagia in MSA needs to be managed by a multidisciplinary team
- Periodic follow-up is required
- No evidence of effective treatment for dysphagia
- Compensatory strategies (i.e. diet modification, swallowing maneuvers and head postures) should be applied when possible
- In specific cases of isolated UES hyperactivity, BTX injections may be effective
- PEG feeding may be applied when there is a severe risk of malnutrition
- No evidence that PEG improves survival/QoL

**Statements on the diagnosis of dysphagia**

Class II to IV studies and on expert adaptation of knowledge from the literature on dysphagia

Patients should be screened at the time of diagnosis and periodically thereafter

- The clinical history should include:
  - Coughing/choking during/after eating or drinking
  - Wet/gurgly voice after eating or drinking
  - Hoarse voice
  - Perception of food or pills stuck in the throat
  - Food modifications or posturing adopted spontaneously
  - Prolonged meal duration
  - Fatigability during meals
  - Drooling or food falling from mouth
  - Recurrent pneumonia
  - Recurrent episodes of fever of unknown origin
  - Unexplained weight loss

- A simple clinical assessment of dysphagia is included in UMSARS.
- The evaluation of dysphagia comprises a screening questionnaire and clinical and instrumental assessment (VFSS, FEES and manometry).
- VFSS and FEES assess both presence and severity of dysphagia
When to assess swallowing in HD?
How to assess dysphagia in HD?
What are the treatments influencing swallowing function in HD?

Guidelines for screening, diagnosis and management of dysphagia in HD are missing.

Systematic review on the assessment and treatment of dysphagia in HD

Management of dysphagia in Huntington’s disease: a descriptive review

What are the treatments influencing swallowing function in HD?
Dysphagia should be **assessed** from the **early stage** of the disease.

Instrumental assessment of swallowing by **VFSS and FEES** is feasible and **recommended** to diagnose dysphagia in HD.

Clinical assessment tools do not replace instrumental assessment but can be complementary.

The impact of **pharmacological and rehabilitative treatments** on dysphagia in HD has been little studied:

- There are no well-proven rehabilitative strategies to improve swallowing function in HD.
Take Home Message…

- Early diagnosis of dysphagia
- Early intervention
- Benefit from the involvement of several professional figures in the management of dysphagia (neurologists, otorhinolaryngologists, phoniatrists, speech and language pathologists and dieticians)
- Need for large, well-designed and longitudinal studies
Thanks …

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