Main Aspects of the Management of Neurogenic Dysphagia

Mario Prosiegel/München

German Society of Neurology (DGN)
prosiegel@t-online.de

DYSPHAGIA
October 8-10, 2015
Pavia, Italy
Overview

Diagnosis

Causal Therapy

General Measures

Symptom-Oriented Therapy
• Rehabilitation
• Invasive Interventions
• Special Conditions

Experimental/New Approaches
Case Report

78 year old woman

- Progressive dysphagia (1 y)
  - Bilateral tongue atrophy with fibrillations
  - Aspiration (food and saliva)

- Fed via PEG
- Admitted for swallowing therapy
- Diagnosis: ALS
A 75 year old woman was admitted to our hospital's swallow- ing department because she had had progressive dysphagia for a year. Based on bilateral tongue atrophy with fibrillations and a normal CT, amyotrophic lateral sclerosis with bulbar onset had been diagnosed 14 months previously, and a percutaneous gastrostomy was performed for feeding purposes.

The neurological examination on admission disclosed a dysphagia with signs of aspirations of saliva, extensive bilateral paresis, and atrophy of the tongue (Fig 1 A) showing diffuse fibrillations. Further neurological examination was normal.

Somatosensory evoked potentials and nerve conduction studies as well as electromyography of various muscles of the upper and lower limbs showed no pathology. Comprehensive laboratory tests and CSF examination produced normal results. A videofluoroscopic swallowing study (VFSS) showed a reduced lingual control, especially for liquid consistencies, with subsequent pharyngeal aspiration. Brain and spinal MRI showed a Chiari I malformation with descent of the cerebellar tonsils to mid-level of the foraminl matrix (Fig 1 B). There were no concomitant malformations such as syringomyelia or arachnoid cysts.

The patient underwent neurosurgical posterior fossa decompression. After the operation, swallowing problems occurred and a tracheostomy was performed due to frequent aspirations of saliva. The patient was readmitted to our hospital with a blocked tracheostomy tube. Oral feeding was not possible. After a 6 week period of swallowing therapy the dysphagia showed a marked improvement. Moisturised partial feeding (about 70%) is possible, the remainder being supplied by the gastrostomy tube. The tracheostomy tube has not yet been removed, as it is required to draw off bronchial mucus, which occasionally causes respiratory problems. A follow-up examination 6 months after the operation showed a bilateral reduction of the atrophy and the fibrillations of the tongue as well as improved tongue function without aspirations on VFS. There were still no signs of a generalised motor neuron disease.

Dysphagia may occur as the sole manifestation of adult Chiari I malformation. As far as we know, there has only been one case published with a Chiari I malformation in an adult mimicking a bulbar palsy in amyotrophic lateral sclerosis. This patient, however, did not show signs of lower motor neuron disease, which were predominant in our patient. We hypothesise that the dysphagia in our patient was caused by pressure exerted by the cerebellar mass on the hypoglossal nuclei and possibly also on the central pattern generators for swallowing situated in the dorsal region of the medulla (Fig 1 C). A Chiari I malformation is an important differential diagnosis in patients with adult onset of dysphagia requiring MRI examination, even when presenting with “typical” lower motor neuron signs in bulbar muscles.

Authors’ affiliations
M Pauly, M Prosiegel
Neurologisches Krankenhaus, Trittaustrasse 20, D-00004 Merseburg, Germany
Correspondence to: Dr M Pauly

REFERENCES
Neurosurgical decompression (removal of part of occipital bone)
Good recovery

## Table 1 Checklist for dysphagia of unknown cause

<table>
<thead>
<tr>
<th>Condition</th>
<th>Tests</th>
</tr>
</thead>
<tbody>
<tr>
<td>CIP/CIM/CIPNM, myotonia, myasthenia gravis, LEMS, GBS</td>
<td>Electromyography, repetitive nerve stimulation, motor and sensory nerve conduction studies</td>
</tr>
<tr>
<td>MS, neuroborreliosis, CPM/EPM, skull base tumors, Chiari malformations</td>
<td>Cranial CT or MRI</td>
</tr>
<tr>
<td>Eagle’s syndrome, ventral osteophytes, and/or complications after anterior cervical spine surgery</td>
<td>Lateral cervical radiography, (3D) CT</td>
</tr>
<tr>
<td>Diseases of the neuromuscular junction</td>
<td></td>
</tr>
<tr>
<td>Myasthenia gravis</td>
<td>Anti-AChR abs, anti-MuSK abs</td>
</tr>
<tr>
<td>LEMS</td>
<td>Anti-VGCC abs</td>
</tr>
<tr>
<td>Myositis</td>
<td>Myositis-associated abs such as</td>
</tr>
<tr>
<td>PM, DM</td>
<td>Anti-Mi-2 abs, anti-SRP abs, antisynthetase (anti-Jo-1) abs</td>
</tr>
<tr>
<td>IBM</td>
<td>Anti-ADDL monoclonal abs</td>
</tr>
<tr>
<td>Connective tissue diseases</td>
<td></td>
</tr>
<tr>
<td>Sjögren’s syndrome</td>
<td>Anti-SS-A/Ro abs, anti-SS-B/La abs</td>
</tr>
<tr>
<td>Systemic sclerosis</td>
<td>Anti-scl70/antitopoisomerase abs; anti-PM-Scl abs</td>
</tr>
<tr>
<td>MCTD/Sharp’s syndrome</td>
<td>Anti-U_{1}-RNP abs</td>
</tr>
<tr>
<td>SLE</td>
<td>Anti-dsDNA abs</td>
</tr>
</tbody>
</table>

From: **Prosiegel M.** Neurology of swallowing and dysphagia.  
### Table 1 continued

<table>
<thead>
<tr>
<th>Vasculitides</th>
<th>ANCA</th>
</tr>
</thead>
<tbody>
<tr>
<td>Wegener’s granulomatosis</td>
<td>Cytoplasmic ANCA (antigen, proteinase 3)</td>
</tr>
<tr>
<td>Microscopic polyangiitis</td>
<td>Perinuclear ANCA (antigen, myeloperoxidase)</td>
</tr>
<tr>
<td>Churg–Strauss syndrome</td>
<td>Perinuclear ANCA (antigen, myeloperoxidase)</td>
</tr>
<tr>
<td>Polyarteritis nodosa</td>
<td>HBsAg (in about 60%)</td>
</tr>
<tr>
<td>Polyneuritis cranialis, Miller–Fisher syndrome</td>
<td>Antiganglioside abs against GQ1b or GT1a</td>
</tr>
<tr>
<td>Paraneoplastic syndromes</td>
<td></td>
</tr>
<tr>
<td>LEMS</td>
<td>Anti-VGCC abs</td>
</tr>
<tr>
<td>Brainstem encephalitis</td>
<td>Anti-Hu, anti-Ri, anti-Ma2 abs</td>
</tr>
<tr>
<td>Stiff-person syndrome</td>
<td>Antiamphiphsin, antigephyrin, anti-Ri abs</td>
</tr>
<tr>
<td>Idiopathic stiff-person syndrome</td>
<td>Anti-GAD abs</td>
</tr>
<tr>
<td>Cerebrospinal fluid examination</td>
<td>Neuroborreliosis, MS, meningitis</td>
</tr>
<tr>
<td>CADASIL</td>
<td>Skin biopsy: granular osmiophilic material in dermal arteries (transmission electron microscopy)</td>
</tr>
<tr>
<td>Myositides, rare myopathies</td>
<td>Muscle biopsy</td>
</tr>
<tr>
<td>CADASIL, SBMA/Kennedy’s disease, OPMD</td>
<td>Molecular genetic examination</td>
</tr>
<tr>
<td>PCNSV/PACNS</td>
<td>Brain biopsy</td>
</tr>
</tbody>
</table>

From: Prosiegel M. Neurology of swallowing and dysphagia.  
Clarify the Pathophysiology – 2 Examples

Example 1
FEES: Predeglutitive pooling in the pharynx
a) Disturbed oral control (tongue, chewing) ? **Solids** > liquids
b) Delayed swallow reflex ? **Liquids** > solids
Treatment (Examples)
a) Tongue strength
b) Chin tuck and/or thickening of liquids

Example 2
FEES: Postdeglutitive residues in the pharynx (bilaterally)
UES opening deficit ?
a) Diminished hyolaryngeal movement ? -> Secondary UES opening deficit
b) Primary UES opening deficit ? (Manometry useful)
Treatment (Examples)
a) Shaker exercise, Mendelsohn maneuver (MM)
b) Shaker exercise, MM, BoNT injection or cricopharyngeal myotomy
Key Statements – Neurogenic Dysphagia – I

Make the **correct diagnosis** as basis for (causal) **therapy**. Doing this, a **checklist** may be helpful.

**Clarify the pathophysiology** (Why and how is swallowing disturbed?).
Diagnosis

Causal Therapy

General Measures

Symptom-Oriented Therapy
• Rehabilitation
• Invasive Interventions
• Special Conditions

Experimental /New Approaches
Dysphagia is necessary, but not sufficient

- Dependent for feeding
- Tube feeding
- Dependent for oral care
- Number of decayed teeth
- More than one medical diagnosis
- Number of medications
- Smoking
Oral Care May Reduce Pneumonia in the Tube-fed Elderly: A Preliminary Study

Keisuke Maeda · Junji Akagi

N = 63

Oral Care Protocol included
- Tooth and tongue brushing
- Oral mucosa brushing
- 0.2% chlorhexidine solution
- Moisturizing the inner mouth with glyceryl polymethacrylate gel
- Salivary gland massage

Fig. 2 a Pneumonia occurrence rate
“The pneumonia rate at sites with a formal dysphagia screen was 2.4% versus 5.4% (p=0.0016) at sites with no formal screen.”
Improving Post-Stroke Dysphagia Outcomes Through a Standardized and Multidisciplinary Protocol: An Exploratory Cohort Study

Mariatalisa Gandolfi · Nicola Smania · Giulia Bisoffi · Teresa Squaquara · Paola Zuccher · Sara Mazzucco

ADMISSION TO THE NEUROLOGY WARD

NEUROLOGIC EVALUATION AND SCREENING OF SWALLOWING DISTURBANCES

YES

Selection of type of nutrition support
NTF/Modified diet

Follow-up

Patients not suitable for rehabilitation:
- TCT score<12/100
- Severe cognitive impairment
- Uncooperative patient

REHABILITATION EVALUATION AND CLINICAL ASSESSMENT OF SWALLOWING
- Head and trunk postural control
- Cooperativeness
- Facial and mouth movements and oral apraxia
- Oropharyngeal structure
- Swallowing trials
- Neurological impairment and disability

Patient suitable for rehabilitation

Rehabilitation Phase I

INSTRUMENTAL EVALUATION
FEES and/or VFSS

Rehabilitation Phase II

Rehabilitation Phase III

Free diet

Rehabilitative phase

Diagnostic phase
Management of patients with stroke: identification and management of dysphagia

June 2010

Can the patient be set up and remain awake and alert for at least 15 minutes?

**YES**

Is the mouth clean?

**YES**

Sit patient up and give a teaspoon of water x 3. Place fingers on midline above and below the larynx and feel the swallow. Observe each teaspoon.

*Are any of these signs present?*
- Absent swallow
- Cough
- Delayed cough
- Altered voice quality

*(ask the patient to say “Aah”)*

**YES**

Keep nil by mouth and refer to Speech and Language Therapist.

**NO**

Implement oral hygiene immediately.

**NO**

Observe the patient continuously drink a glass of water.

*Are any of these signs present?*
- Absent swallow
- Cough
- Delayed cough
- Altered voice quality

*(ask the patient to say “Aah”)*

**YES**

Keep nil by mouth and refer to Speech and Language Therapist.

**NO**

Start feeding (soft options) with caution.

Continue to observe for coughing or development of a chest infection and refer to Speech and Language Therapy as necessary.
Key Statements – Neurogenic Dysphagia – I

Make the **correct diagnosis** as basis for (causal) **therapy**. Doing this, a **checklist** may be helpful.

**Clarify the pathophysiology** (Why and how is swallowing disturbed?).

*Perform oral care and use a screening procedure/algorithm to prevent/reduce aspiration pneumonia in acute stroke.*
Diagnosis

Causal Therapy

General Measures

**Symptom-related Therapy**
- **Rehabilitation**
- Invasive Interventions
- Special Conditions

Experimental /New Approaches
Rehabilitation

Restitution (indirect methods)
improves disturbed functions (by promoting neuroplasticity)
(Use it or lose it! Use it and improve it!)
• Exercises (e.g. Shaker/head-rising exercise)

Compensation (direct methods)
improves swallowing despite disturbed functions (temporarily)
• Posture changes (e.g. head turning)
• Maneuvers (e.g. Mendelsohn, supraglottic swallowing)

Adaptation
changes the environment, e.g.
• Consistency modification (e.g. thickening liquids)
• Texture modification (e.g. avoiding sticky rice cakes)
Efficacy of exercises to rehabilitate dysphagia: A critique of the literature
SUSAN E. LANGMORE¹,² & JESSICA M. PISEGNA²

Evidence for long-term benefit
– Randomized Controlled Trials (RCTs) –

• Shaker exercise
  (N=72; Shaker et al., 1997 & 2002; Logemann et al., 2009)

• Expiratory Muscle Strength Training (EMST) in PD
  (N=60; Troche et al., 2010)

• Mendelsohn maneuver
  (N=18; McCullough et al., 2012)
**Shaker exercise**

From: Mepani R et al.
Dysphagia 2009;24:26-31

**EMST**

Calibrated pressure scale

Mouthpiece

One-way valve

From: Pitts T et al.
Chest 2009;135:1301-8

**Mendelsohn maneuver (EMG Biofeedback)**

From: www.bfe.org/protocol/pro06eng.htm

*Figure 7. EMG Tracing of Mendelsohn Maneuver*
## Restitutional / Indirect interventions

<table>
<thead>
<tr>
<th>Disturbance</th>
<th>Intervention</th>
<th>Mechanism</th>
</tr>
</thead>
<tbody>
<tr>
<td>UES dysfunction</td>
<td>Head-rising (Shaker) exercise</td>
<td>Strengthening of suprathyoidal muscles</td>
</tr>
<tr>
<td>Dysphagia in Parkinson disease</td>
<td>Expiratory muscle strength training (EMST)</td>
<td>Strengthening of expiratory and suprathyoidal muscles</td>
</tr>
<tr>
<td>Reduced tongue force/pharyngeal transit</td>
<td>Tongue-holding (Masako) exercise</td>
<td>Improving BOT to PPW approximation</td>
</tr>
<tr>
<td>Dysphagia in Parkinson disease</td>
<td>Lee-Silverman-Voice-Treatment® (LSVT®)</td>
<td>Improving voice and swallowing parameters</td>
</tr>
</tbody>
</table>
## Compensatory / Direct interventions

<table>
<thead>
<tr>
<th>Disturbance</th>
<th>Intervention</th>
<th>Mechanism</th>
</tr>
</thead>
<tbody>
<tr>
<td>UES dysfunction</td>
<td>Mendelsohn maneuver</td>
<td>Enhancing width of UES/duration of UES opening</td>
</tr>
<tr>
<td>Predeglutititive or deglutititative aspiration</td>
<td>Supraglottic swallowing</td>
<td>Closing of vocal folds and clearing of laryngeal residues</td>
</tr>
<tr>
<td>Predeglutitive aspiration</td>
<td>Chin tuck</td>
<td>Narrowing of laryngeal entrance</td>
</tr>
<tr>
<td>Unilateral pharynx paresis</td>
<td>Head turning to ipsilateral side</td>
<td>Directing bolus down the stronger side</td>
</tr>
</tbody>
</table>
Predeglutitive Aspiration
Leaking due to delayed swallow reflex
Solid bolus

Premature spillage due to insufficient lingual control
27 studies (from 10 147 !)

“… thicker liquids reduce the risk of penetration/aspiration, but also increase the risk of post-swallow residue in the pharynx.“
N = 20 healthy participants

„The results showed increased velocities and higher peak velocities with the nectar-thick stimuli compared to thin and ultrathin stimuli.”

Nectar-thick: 236 mPa s @ 50 reciprocal seconds
Utilizing thin, thick and ultra-thick liquids and delivery by cup and spoon increases the chance of finding a consistency that the patient can swallow without aspiration or pharyngeal residues (> 90% of patients with mild or moderate dysphagia).
Swallowing disorders in tracheostomised patients: a multidisciplinary/multiprofessional approach in decannulation protocols

Giancarlo Garuti, Cristina Reverberi, Angelo Briganti, Monica Massobrio, Francesco Lombardi and Mirco Lusuardi

Dysphagia evaluation in Tracheostomized patients

1. Secretion inhalation
2. Desaturation episodes
3. Respiratory complications

STOP decannulation protocol
Maintenance of cuffed cannula
No decannulation

DYSPHAGIC PATIENT
Postpone decannulation to potential dysphagia resolution

1. No secretion inhalation
2. No desaturation episodes
3. No respiratory complications

Swallowing Test (Fluids, semisolid and solid foods)

NON DYSPHAGIC PATIENT
Decannulation
Assessment of Outcome

Validated Scales

Body Function or Structure, e.g.
- VFSS, FEES, Manometry
- Aspiration pneumonia, mortality
- PAS (Penetration-Aspiration Scale)

Activity, e.g.
- DOSS (Dysphagia Outcome and Severity Scale)

Participation, e.g.
- QoL (SWAL-QOL)

ICF (WHO)
International Classification of Functioning, Disability and Health
Key Statements – Neurogenic Dysphagia – I

Make the **correct diagnosis** as basis for (causal) **therapy**. Doing this, a **checklist** may be helpful.

**Clarify the pathophysiology** (Why and how is swallowing disturbed?).

Perform **oral care** and use a **screening procedure/algorithm** to prevent/reduce aspiration pneumonia in acute stroke.

*Use symptom-oriented and **evidence-based rehabilitation measures**. Good evidence exists, e.g., for Shaker exercise, EMST, Mendelsohn maneuver and increasing bolus viscosity. When removal of a **tracheal cannula** is planned, use a **decannulation protocol**.

**Check the efficacy** of posture changes, maneuvers and consistency/texture modifications by use of **FEES** (or **VFSS**) in individual patients under **different conditions**.

*Use **validated outcome scales** for assessing **ICF components**.*
Diagnosis

Causal Therapy

General Measures

**Symptom-related Therapy**
- Rehabilitation
- **Invasive Interventions**
- Special Conditions

Experimental /New Approaches
Dysfunction of the Upper Esophageal Sphincter


Botulinum toxin for upper oesophageal sphincter dysfunction in neurological swallowing disorders (Review)

Regan J, Murphy A, Chiang M, McMahon BP, Coughlan T, Walshe M

Comments

Currently, no evidence is available to support the routine use of botulinum toxin to treat neurogenic dysphagia. Methodologically sound randomised controlled trials are urgently required in order to verify its safety and clinical value across various adult neurogenic groups and to determine optimal candidacy and protocols.
The question is open, whether **botulinum toxin injection** is superior to **myotomy**. Although further studies are needed, you must help patients individually dependent on the existing **special expertise** in your institution.
Diagnosis

Causal Therapy

General Measures

**Symptom-related Therapy**
- Rehabilitation
- Invasive Interventions
- **Special Conditions**

Experimental /New Approaches
Three Examples of Special Problems

1) Diseases with (progressive) muscle weakness, e.g. ALS, Myasthenia gravis, Myositis, Post-Polio Syndrome
   – Avoid exhaustion
   – Avoid distraction
   – ‘Little and often’ approach (meals, therapy)

2) Cognitive deficits, Old patients
   – Avoid distraction
   – Use simple instructions
   – Assess nutritional status

3) Hypersalivation/Sialorrhea, e.g. PD, Severe dysphagia
   – Pharmacological reduction of saliva
Diagnosis

Causal Therapy

General Measures

Symptom-Oriented Therapy
• Rehabilitation
• Invasive Interventions
• Special Conditions

Experimental/New Approaches
Role of Cerebral Cortex Plasticity in the Recovery of Swallowing Function Following Dysphagic Stroke

Andrew W. Barritt · David G. Smithard

rTMS (repetitive Transcranial Magnetic Stimulation)
- excitatory (3-5 Hz)

tDCS (transcranial Direct Current Stimulation)
- excitatory (anodal)

PES (Pharyngeal Electrical Stimulation)
- 5 Hz
3 rTMS and 3 tDCS studies

Statistically significant beneficial effect of
• rTMS compared with sham stimulation
• not of tDCS

Ipsilesional vs. contralesional site of stimulation
• no statistically significant difference
The question is open, whether botulinum toxin injection is superior to myotomy. Although further studies are needed, you must help patients individually dependent on the existing special expertise in your institution.

In certain conditions, special interventions are needed. Examples are a) diseases with (progressive) muscle weakness (ALS, myasthenia gravis etc.), b) cognitive deficits/old persons and c) sialorrhea (e.g. in Parkinson disease). You should avoid muscle exhaustion (a) and distraction (b), use simple instructions (b), reduce saliva production (pharmacologically) (c) and assess the nutritional status.

Examples of new interventions comprise PES, rTMS and tDCS.