Addressing Respiratory Challenges in ALS

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Addressing Respiratory Challenges in ALS

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Disclosures

- I do NOT have any relevant financial relationships with any commercial interests.
- Teach in RT degree advancement (DA) programming (BS & MS)
- Board member, Commission on Accreditation for Respiratory Care (CoARC)
- Voluntary staff therapist at a multidisciplinary ALS clinic since 2016
Objectives

As a result of attending today’s webinar, the learner will be able to:

- Identify how ALS impacts the respiratory system
- Discuss options currently available to help aid in the respiratory management of the pALS
- Approach the multidisciplinary team with questions and preferences regarding their respiratory care
How ALS presents itself

- Affects motor nerve cells in brain and spinal cord which results in loss of muscle control
  - Speaking
  - Swallowing
  - Walking
  - Grasping
  - **Breathing**
- **Limb Onset**
  - Trouble grasping with hands
  - Tripping, dropping things
- **Bulbar Onset**
  - Tongue fasciculations
  - Dysphagia, sialorrhea

Respiratory Challenges

https://www.als.org/understanding-als/what-is-als
Respiratory Involvement

- Less than 3% of cases present with respiratory symptom onset initially
- Respiratory involvement eventually occurs in all patients with ALS
- Pulmonary complications are a heightened concern in the disease process
- Weak muscles of the diaphragm and muscles of respiration result in
  - Difficulty inhaling sufficient air with each breath
  - Difficulty clearing the airway with a strong cough
- Respiratory symptoms include poor sleep quality, low oxygen levels (especially during sleep), difficulty breathing while lying flat, excessive daytime sleepiness, headaches, rapid/shallow breathing, and shortness of breath with or without exertion
Goals of Care when Respiratory System is Affected

1. Prevent chest/respiratory infections (through appropriate vaccinations)
1. Decrease work of breathing (consider noninvasive ventilation)
1. Maintain a patent airway (manually assisted cough, mechanical assisted cough, secretion management, suction machine, or invasive ventilation)
1. Treat any infections or respiratory insufficiency/failure as it occurs
1. Enhance survivability and maximize quality of life as able
What guides our respiratory recommendations?


1. Discussion with our pALS concerning their preferences for care*
AAN: Respiratory Management Algorithm

Key Components of Algorithm:

- PFT/spirometry every 3-6 months
  - Multidisciplinary clinic preferred
- Airway clearance strategies
  - Manual v. mechanical
- Secretion/sputum management
  - Suction, medications
- Measure respiratory muscle strength
  - MIP, SNP, MEP
- Ventilation
  - NIV or invasive, overnight oximetry
Respiratory Evaluation

**Need for ventilation assistance:**
- Orthopnea (difficulty lying flat)
- Sniffing nasal pressure (< 40 cm H2O)
- Maximal inspiratory pressure (< -60 cmH2O)
- Abnormal oxygen saturation during sleep
- Forced (or slow) vital capacity < 50% of predicted values (sitting or supine)
- PaCO2 > 45 mmHg

**Need for cough assistance:**
- Peak expiratory cough flow < 270 L/m
- Maximum expiratory pressure < 40 cmH2O

Respiratory Challenges
Respiratory Monitoring

**Forced vital capacity (FVC)**
- Maximum amount of air that can be exhaled as quickly and forcefully as possible after maximum inspiration
- Based on age, gender, height, ethnicity, and weight

**Maximum inspiratory pressure (MIP)**
- Maximum amount of negative pressure generated during inspiration through an occluded system
- Decline in MIP may occur 4-6 months before FVC

Respiratory Challenges
Respiratory Monitoring

**Sniff nasal pressure**
- Noninvasive means of measuring inspiratory muscle strength
- May be easier for some patients when compared to MIP
- Correlates to decline in FVC and hypercapnia

**Maximum expiratory pressure**
- Highest positive pressure that can be generated during a forced expiration against an occluded airway
Cough for Airway Clearance & Protection

Respiratory Challenges

https://clinicalgate.com/airway-clearance-therapy/
Manual Cough Assistance Options

Abdominal thrust maneuver

Glossopharyngeal “frog” breathing

Mechanical Cough Assistance Options

- Mechanical insufflation-exsufflation (MIE) or “Cough Assist”
- Inspiratory and expiratory pressures are set based on patient tolerance and clearability of proximal secretions
- Generally 20-40 cmH2O
- Helpful in the presence of pulmonary infections
- May reduce morbidity and hospitalization
- May be less effective in those with bulbar involvement

Respiratory Challenges
Managing Secretions

**Dysphagia** *(difficulty swallowing)*

- Restricted tongue movement/mouth opening, bulbar muscle atrophy, and laryngeal dysfunction are all characteristics of advanced dysphagia in ALS
- Altered respiratory-swallow phase pattern in subjects with ALS with higher rates of non-ideal exhalation surrounding the swallowing movement

**Sialorrhea** *(excessive oral secretions)*

- Average salivary production is roughly 0.5-1.5 L/day and any loss of clearability can fall within a spectrum of mild inconvenience to respiratory distress depending on the degree of overwhelm
- Oral Secretion Scale (OSS) can be used at the bedside to measure secretions relative to the individual’s ability to swallow

**Respiratory Challenges**
## Oral Secretion Scale

<table>
<thead>
<tr>
<th>OSS Score</th>
<th>Oral Secretion Characteristics</th>
<th>Saliva Swallow Ability</th>
</tr>
</thead>
<tbody>
<tr>
<td>4, Normal</td>
<td>No excessive secretions</td>
<td>Automatic, normal</td>
</tr>
<tr>
<td>3, Mild</td>
<td>Infrequent, small accumulation of secretions in the mouth; infrequent wet lips or drooling; infrequent lip blotting</td>
<td>Automatic, decreased</td>
</tr>
<tr>
<td>2, Moderate</td>
<td>Occasional drooling, lip blotting; occasional pooling of secretions in the throat; oropharyngeal suctioning 0–2/h</td>
<td>Conscious, required</td>
</tr>
<tr>
<td>1, Severe</td>
<td>Frequent drooling, lip blotting; frequent pooling of secretions in the throat; oropharyngeal suctioning 3–4/h</td>
<td>Conscious, difficult</td>
</tr>
<tr>
<td>0, Most severe</td>
<td>Constant drooling, lip blotting; constant pooling of secretions in the throat; oropharyngeal suctioning &gt;4/h</td>
<td>Conscious, impossible</td>
</tr>
</tbody>
</table>

The observer chooses a single score whose criteria most closely match the patient’s condition, according to the worst performance; if multiple criteria in multiple score categories are observed, then the observer selects the score with the most criteria that match the patient’s worst performance; if the observer is uncertain whether the patient has an OSS score of 3 or 2 or an OSS score of 1 or 0, the observer chooses the worst score as the default.
## Managing Secretions

**TABLE 4** Recommended Therapies for Sialorrhea

<table>
<thead>
<tr>
<th>Therapy</th>
<th>Suggestions</th>
<th>Remarks</th>
</tr>
</thead>
<tbody>
<tr>
<td>Anticholinergic medications</td>
<td>• An initial trial of an inexpensive oral anticholinergic is suggested.</td>
<td>• Relatively inexpensive and readily available.</td>
</tr>
<tr>
<td></td>
<td>• Continue to use if the benefits are greater than the side effects.</td>
<td>• Individual patient benefits and adverse events can be assessed easily.</td>
</tr>
<tr>
<td></td>
<td>• More expensive and potentially longer-acting anticholinergic patch medication also can be considered.</td>
<td></td>
</tr>
<tr>
<td>Botulinum toxin therapy to salivary glands</td>
<td>• Limited data, doses are not defined.</td>
<td>• Inexpensive, lasting beneficial effects on salivary function.</td>
</tr>
<tr>
<td></td>
<td>• See individual studies for doses in e-Table 8b.</td>
<td>• May need to be repeated. Associated with viscous saliva and mild to moderate pain.</td>
</tr>
<tr>
<td>Salivary gland RT</td>
<td>• Limited data, doses not defined.</td>
<td>• Long-lasting relief; however, associated with irreversible dryness.</td>
</tr>
<tr>
<td></td>
<td>• See individual studies for doses e-Table 8c.</td>
<td>• Suggest reserving RT to experienced centers.</td>
</tr>
</tbody>
</table>

RT = radiation therapy.
Managing Secretions

**Anticholinergics:**
- Glycopyrrolate, hyoscyamine, scopolamine patch, atropine drops

**Radiation therapy or botulinum toxin injection**

**Surgery:**
- Total laryngectomy
Initiation of Noninvasive Ventilation

- Early use has shown to increase mean survival rate (before FVC < 50%)
- Goal of at least four hours per day

- Use of ventilation support may not stop the progression of ALS, but can
  - Increase energy
  - Improve dyspnea
  - Greater sleep quality
  - Enhanced concentration
  - Decrease fatigue/depression
  - Lengthen survival
  - Slow rate of FVC decline
  - Enhance QoL

Conduct Pulmonary Function Testing (PFT)

- Do results include any of the following?
  - FVC <80% predicted
  - MIP <60 cm H2O or MEP <40 cm H2O
  - PCF <270 L/min
  - SNIP <70 cm H2O (males) or <60 cm H2O (females)

Conduct Overnight Oximetry or ABG Testing

- Do results include any of the following?
  - SpO2 \( \leq 90\% \) for \( \geq 2\% \) sleep time
  - PaCO2 on ABG > 45 mmHg

Conduct Polysomnography

- Do results include any of the following?
  - AHI \( \geq 5\)
  - SpO2 \( \leq 88\% \) for \( \geq 5\) minutes continuously

Repeat PFT in 6 months
NIV Interface Options

Nasal Mask

Full Face Mask
Hybrid

Respiratory Challenges
NIV Interface Options

Mouthpiece Ventilation, Sometimes called “sip and puff”
NIV Compliance/Tolerance

- Bulbar involvement = decreased tolerance to NIV
- Cognition level of patient and characteristics of caregiver
- Participation in multidisciplinary clinic with respiratory/pulmonary involvement has been shown to increase use of/access to NIV

Respiratory Challenges

Additional Treatment Options

If NIV is declined or not tolerated, discuss other treatment options:

- Tracheostomy with invasive ventilation (TIV)
- Discuss withdrawal conditions if TIV is accepted
- Personal, financial, cultural factors leading to the decision
- Ventilation settings should be evaluated frequently and adjusted as needed
Invasive Ventilation

Respiratory Challenges
Palliative care is available at the point of diagnosis. Hospice care is generally sought toward the end of life. Palliative care focuses on support, while hospice care focuses on comfort. Hospice & Palliative Care: How do they compare?
Multidisciplinary Approach

*Also work closely with Neurologists/Pulmonologists, ALS Association Reps, Equipment Specialists

Physical & Occupational Therapists
Registered Dietitian & Pharmacist
Speech Language Pathologist & Social Worker
In-Home Providers

Respiratory Challenges
# ALS Functional Rating Scale

## Dyspnea

<table>
<thead>
<tr>
<th>None</th>
<th>+4</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
<td></td>
</tr>
<tr>
<td>Occurs when walking</td>
<td>+3</td>
</tr>
<tr>
<td>Occurs with one or more of the following: eating, bathing, dressing</td>
<td>+2</td>
</tr>
<tr>
<td>Occurs at rest, difficulty breathing when either sitting or lying</td>
<td>+1</td>
</tr>
<tr>
<td>Significant difficulty, considering using mechanical respiratory support</td>
<td>0</td>
</tr>
</tbody>
</table>

## Orthopnea

<table>
<thead>
<tr>
<th>None</th>
<th>+4</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
<td></td>
</tr>
<tr>
<td>Some difficulty sleeping at night due to shortness of breath; does not routinely use &gt;2 pillows</td>
<td>+3</td>
</tr>
<tr>
<td>Needs extra pillows in order to sleep (&gt;2)</td>
<td>+2</td>
</tr>
<tr>
<td>Can only sleep sitting up</td>
<td>+1</td>
</tr>
<tr>
<td>Unable to sleep</td>
<td>0</td>
</tr>
</tbody>
</table>

## Respiratory Insufficiency

<table>
<thead>
<tr>
<th>None</th>
<th>+4</th>
</tr>
</thead>
<tbody>
<tr>
<td>None</td>
<td></td>
</tr>
<tr>
<td>Intermittent use of BiPAP</td>
<td>+3</td>
</tr>
<tr>
<td>Continuous use of BiPAP during the night</td>
<td>+2</td>
</tr>
<tr>
<td>Continuous use of BiPAP during the night and day</td>
<td>+1</td>
</tr>
<tr>
<td>Invasive mechanical ventilation by intubation or tracheostomy</td>
<td>0</td>
</tr>
</tbody>
</table>

### Respiratory Challenges

Questions to Ask your RT

- Know your numbers! Do the results of pulmonary function testing indication respiratory involvement?
- What airway clearance strategies are ideal for my current status?
- What secretion management options are available for me?
- What type of ventilation support is right for me?
References


Cazzolli, PA et al. (2020). The Oral Secretion Scale and Prognostic Factors for Survival in Subjects With Amyotrophic Lateral Sclerosis. Respiratory Care, 65(8), 1063-1076. https://doi.org/10.4187/respcare.07005


Respiratory Challenges
Thank you for your time and attention!

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