



WELCOME!

September 18, 2023

Addressing Respiratory Challenges in ALS

Guest Speaker:

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Living with ALS Webinar Series:
September 18, 2023

Addressing Respiratory Challenges in ALS

Presenter: Kristen McHenry EdD, RRT-ACCS

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

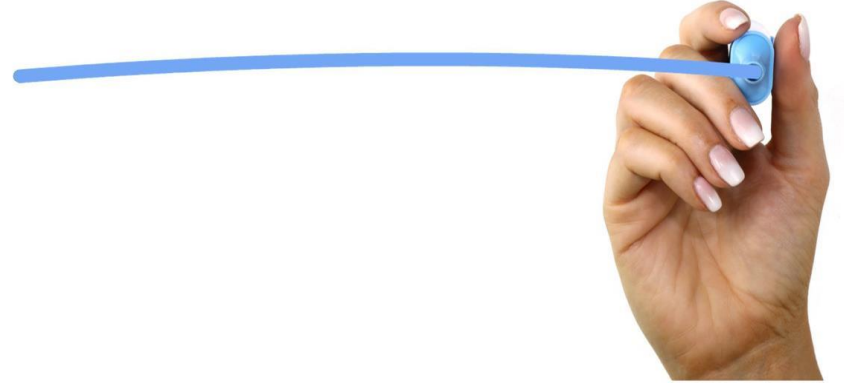


Objectives

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

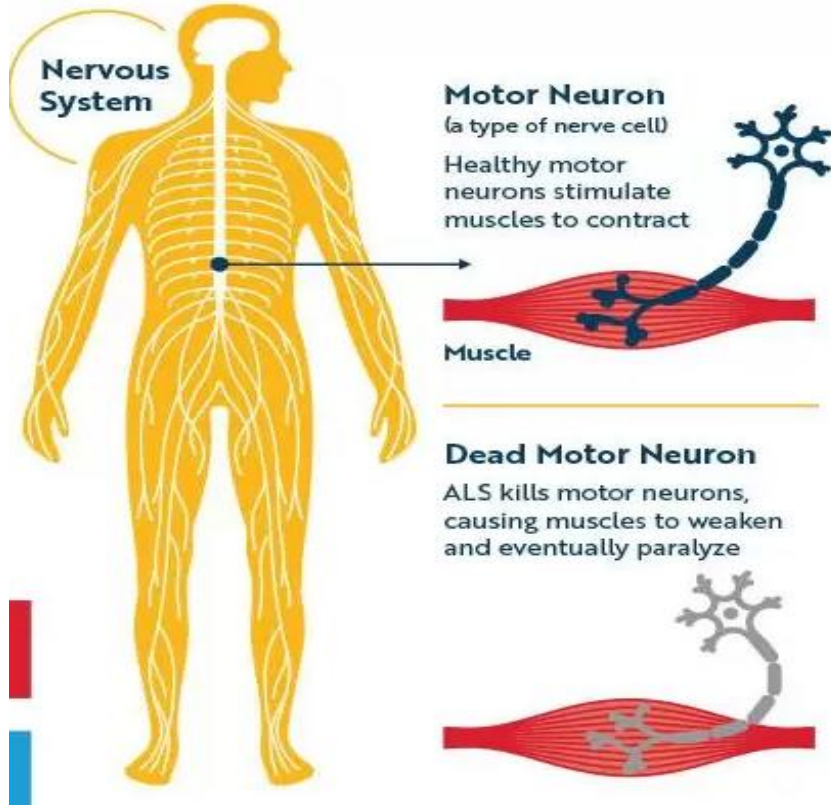
- Identify how ALS impacts the respiratory system
- Discuss options currently available to help aid in the respiratory management of the pALS
- Approach the multidisciplinary team with questions and preferences regarding their respiratory care

OBJECTIVES



Respiratory Challenges

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

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?

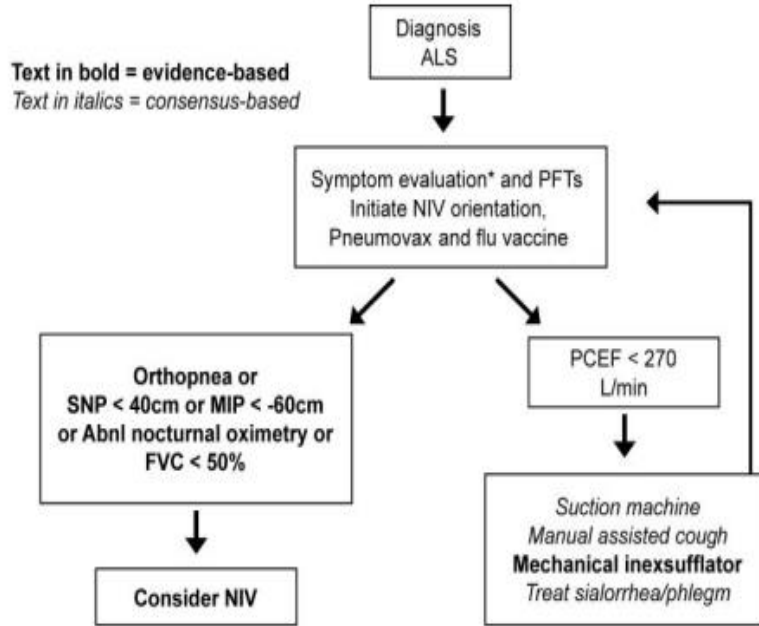
1. Practice Parameter update: The care of the patient with amyotrophic lateral sclerosis: Drug, nutritional, and **respiratory therapies** (an evidence-based review). 2009
1. **Respiratory Management** of Patients With Neuromuscular Weakness: An American College of Chest Physicians Clinical Practice Guideline and Expert Panel Report. 2023
1. Discussion with our pALS concerning their preferences for care*



Respiratory Challenges

AAN: Respiratory Management Algorithm

Figure 2 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 Challenges

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

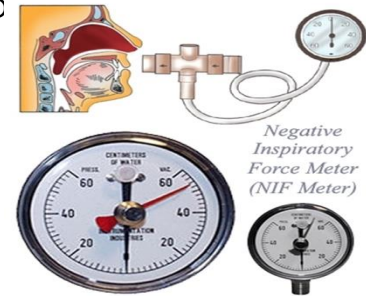
Forced vital capacity (FVC)

- Maximum amount of air that 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 befo



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

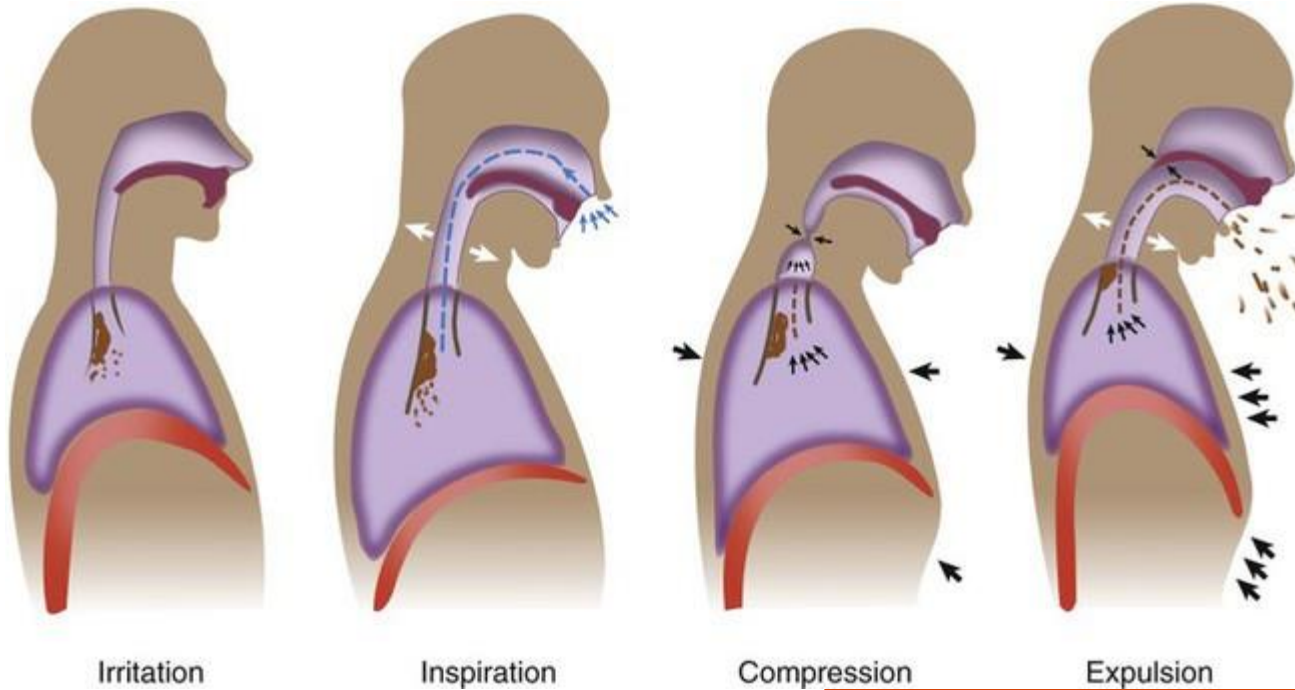
Maximum expiratory pressure

- Highest positive pressure that can be generated during a forced expiration against an occluded airway



Respiratory Challenges

Cough for Airway Clearance & Protection



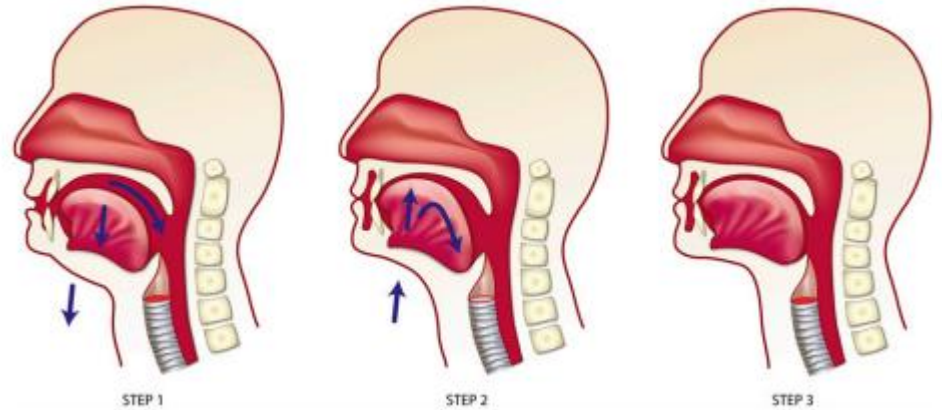
Respiratory Challenges

Manual Cough Assistance Options



Abdominal thrust maneuver

Glossopharyngeal “frog” breathing



Respiratory Challenges

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

Oral Secretion Scale

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

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

Therapy	Suggestions	Remarks
Anticholinergic medications	<ul style="list-style-type: none">• An initial trial of an inexpensive oral anticholinergic is suggested.• Continue to use if the benefits are greater than the side effects.• More expensive and potentially longer-acting anticholinergic patch medication also can be considered.	<ul style="list-style-type: none">• Relatively inexpensive and readily available.• Individual patient benefits and adverse events can be assessed easily.
Botulinum toxin therapy to salivary glands	<ul style="list-style-type: none">• Limited data, doses are not defined.• See individual studies for doses in e-Table 8b.	<ul style="list-style-type: none">• Inexpensive, lasting beneficial effects on salivary function.• May need to be repeated. Associated with viscous saliva and mild to moderate pain.
Salivary gland RT	<ul style="list-style-type: none">• Limited data, doses not defined.• See individual studies for doses e-Table 8c.	<ul style="list-style-type: none">• Long-lasting relief; however, associated with irreversible dryness.• Suggest reserving RT to experienced centers.

RT = radiation therapy.

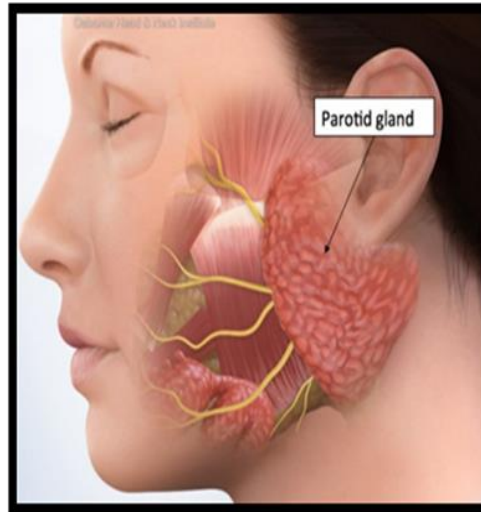
Respiratory Challenges



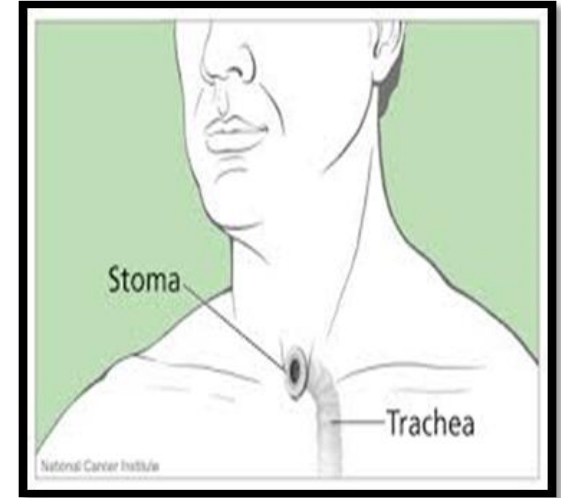
Managing Secretions



Anticholinergics:
Glycopyrrolate,
hyoscyamine,
scopolamine patch,
atropine drops



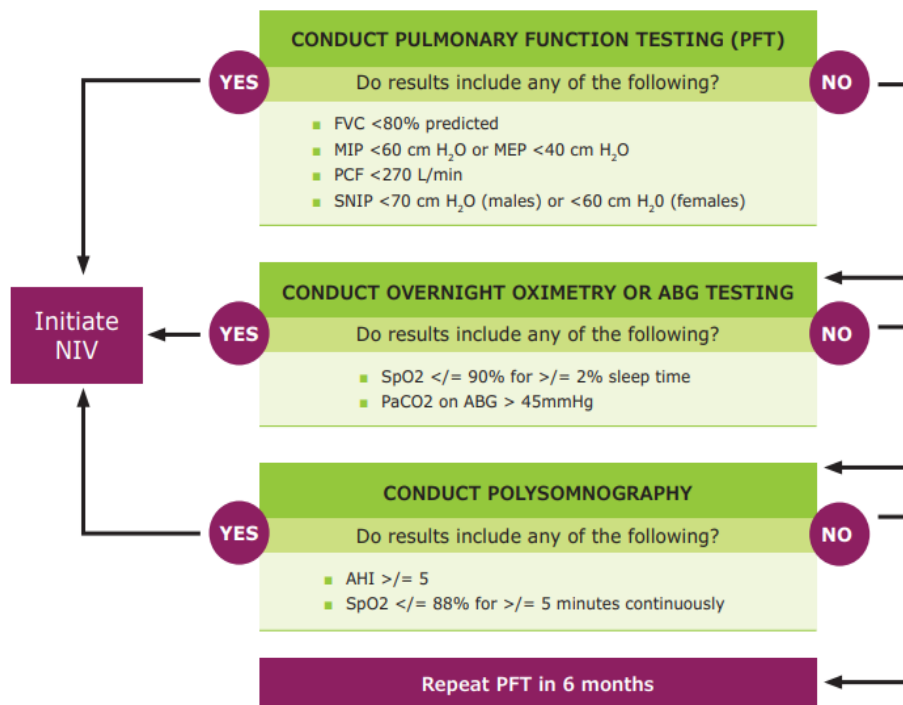
Radiation therapy or
botulinum toxin injection



Surgery:
Total laryngectomy

Respiratory Challenges

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

Respiratory Challenges

NIV Interface Options



Nasal Mask



**Full Face Mask
Hybrid**



Respiratory Challenges

NIV Interface Options

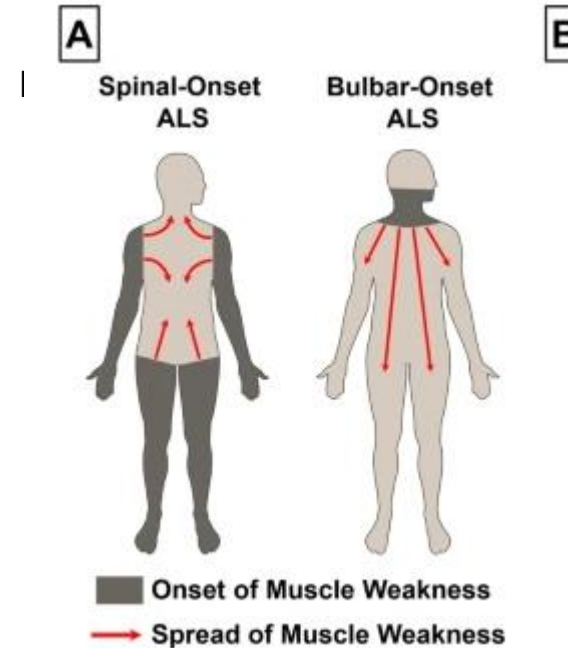


Mouthpiece Ventilation, Sometimes called “sip and puff”

Respiratory Challenges

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

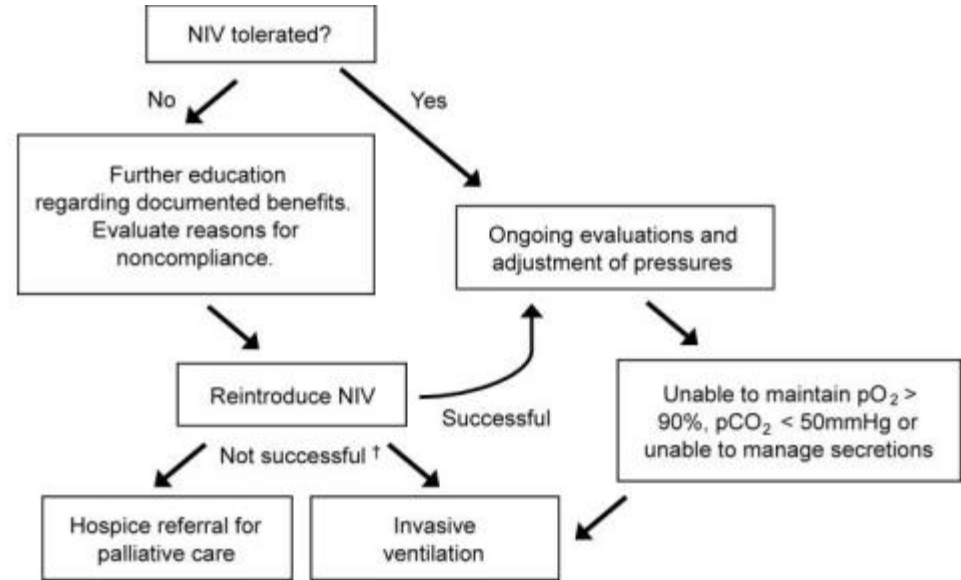


Respiratory Challenges

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

Hospice & Palliative Care



Respiratory Challenges



Multidisciplinary Approach



Physical &
Occupational
Therapists



Registered Dietitian
& Pharmacist



Speech Language
Pathologist & Social
Worker



In-Home Providers

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

Respiratory Challenges

ALS Functional Rating Scale

Dyspnea

Respiratory Insufficiency

None	+4
Occurs when walking	+3
Occurs with one or more of the following: eating, bathing, dressing	+2
Occurs at rest, difficulty breathing when either sitting or lying	+1
Significant difficulty, considering using mechanical respiratory support	0

Orthopnea

None	+4
Some difficulty sleeping at night due to shortness of breath; does not routinely use >2 pillows	+3
Needs extra pillows in order to sleep (>2)	+2
Can only sleep sitting up	+1
Unable to sleep	0

None	+4
Intermittent use of BiPAP	+3
Continuous use of BiPAP during the night	+2
Continuous use of BiPAP during the night and day	+1
Invasive mechanical ventilation by intubation or tracheostomy	0

Respiratory Challenges



Questions to Ask your RT

- Know your numbers! Do the results of pulmonary function testing indicate 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

ALS Association. (2023). *Understanding ALS*. <https://www.als.org/understanding-als/what-is-als>


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>

I am ALS. (2023). *Palliative and hospice care for ALS*. <https://iamals.org/get-help/palliative-and-hospice-care-for-als/#:~:text=Palliative%20care%20is%20available%20to,are%20no%20longer%20seeking%20treatment.>

Khan, A et al. (2023). Respiratory Management of Patients With Neuromuscular Weakness: An American College of Chest Physicians Clinical Practice Guideline and Expert Panel Report. *CHEST*, 164(2), 394-413. <https://doi.org/10.1016/j.chest.2023.03.011>

MD Calc. (2023). *Revised Amyotrophic Lateral Sclerosis Functional Rating Scale (ALSFRS-R)*. <https://www.mdcalc.com/calc/10166/revised-amyotrophic-lateral-sclerosis-functional-rating-scale-alsfrs-r>

Miller, RG et al. (2009). Practice Parameter update: The care of the patient with amyotrophic lateral sclerosis: Drug, nutritional, and respiratory therapies (an evidence-based review). *Neurology*, 73(15), 1218-1226. <https://doi.org/10.1212/WNL.0b013e3181bc0141>



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Thank you for your
time and attention!