Recognizing Deterioration in Neuromuscular Disease:

The Role of Ventilatory Support

Christal M. Hawkins, RRT, BSRT
Respiratory Educator & Ambulatory Supervisor
UC San Diego Health

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No relevant financial or research relationships to disclose

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Learning Objectives

By the end of this session, participants will be able to:

1

Recognize early signs of respiratory deterioration in neuromuscular disease.

2

Identify indicators of ineffective noninvasive ventilation.

3

Explain how ventilator optimization affects patient outcomes.

4

Apply a practical approach to NIV optimization.

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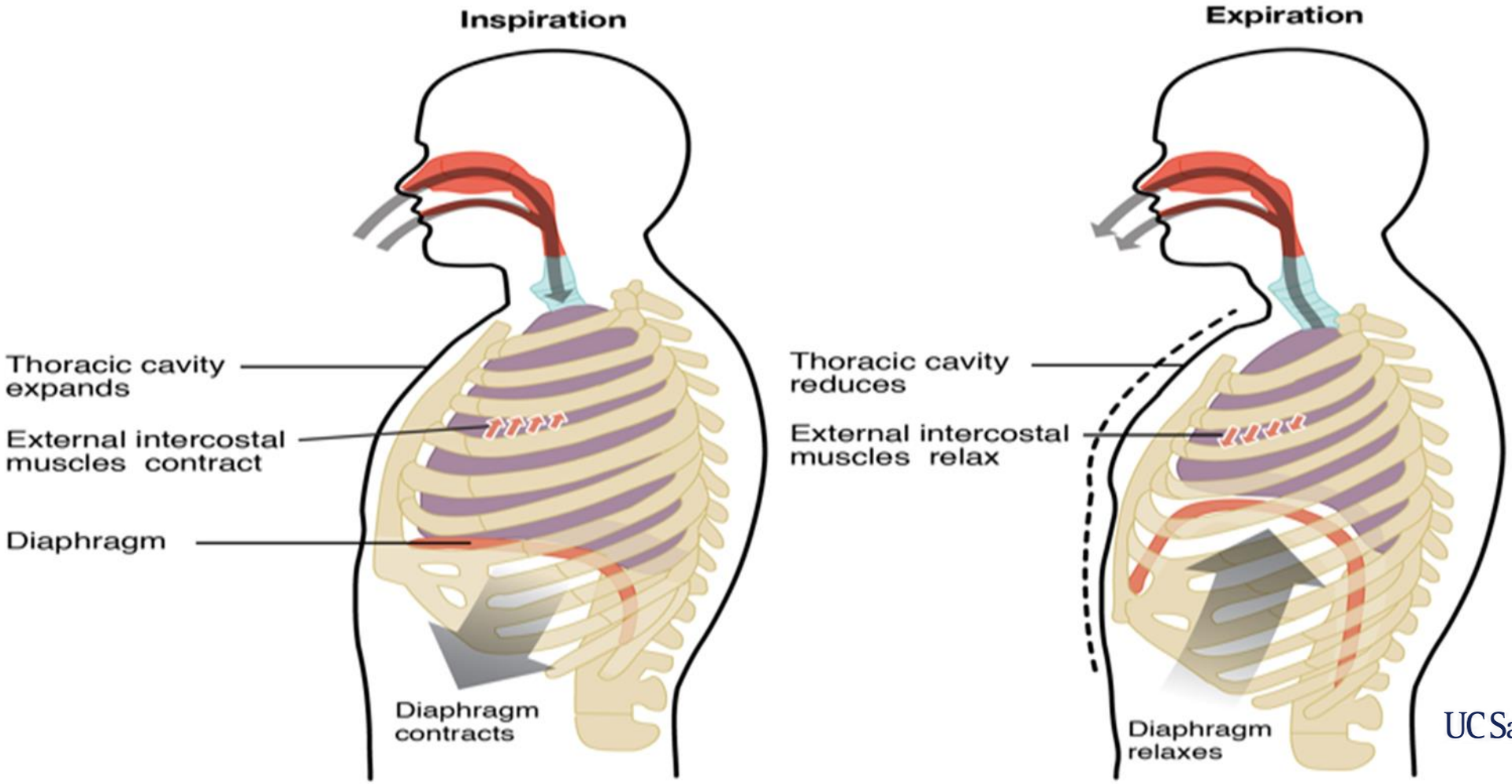
SICK MUSCLES



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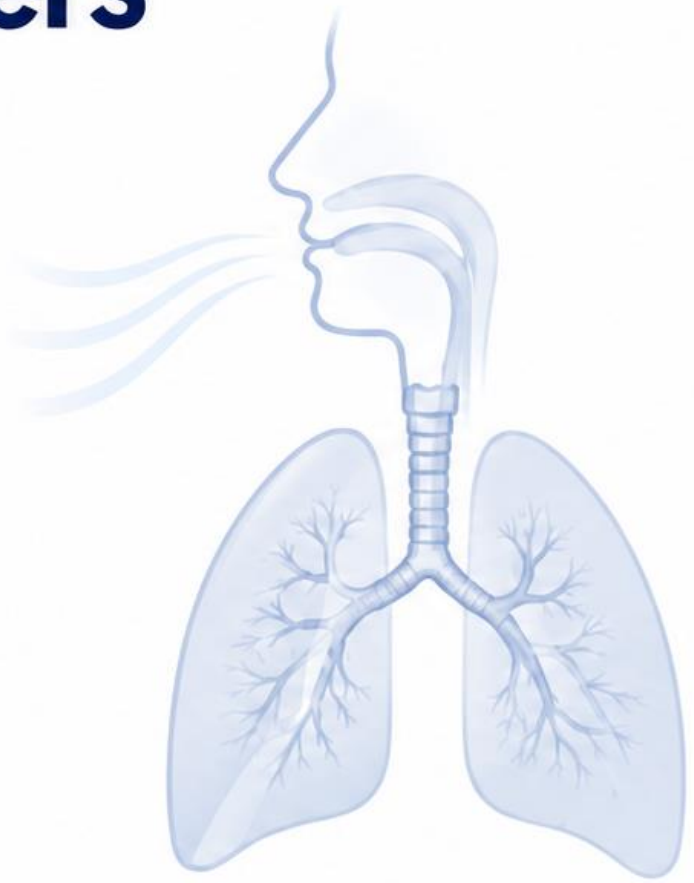
Why Early Recognition Matters

Respiratory decline in neuromuscular disease is often advanced before it becomes clinically obvious.

Subtle.
Easily missed.

Progressive.
Often silent.

Delayed recognition
increases risk.



Early recognition preserves opportunities, reduces complications, and improves outcomes.

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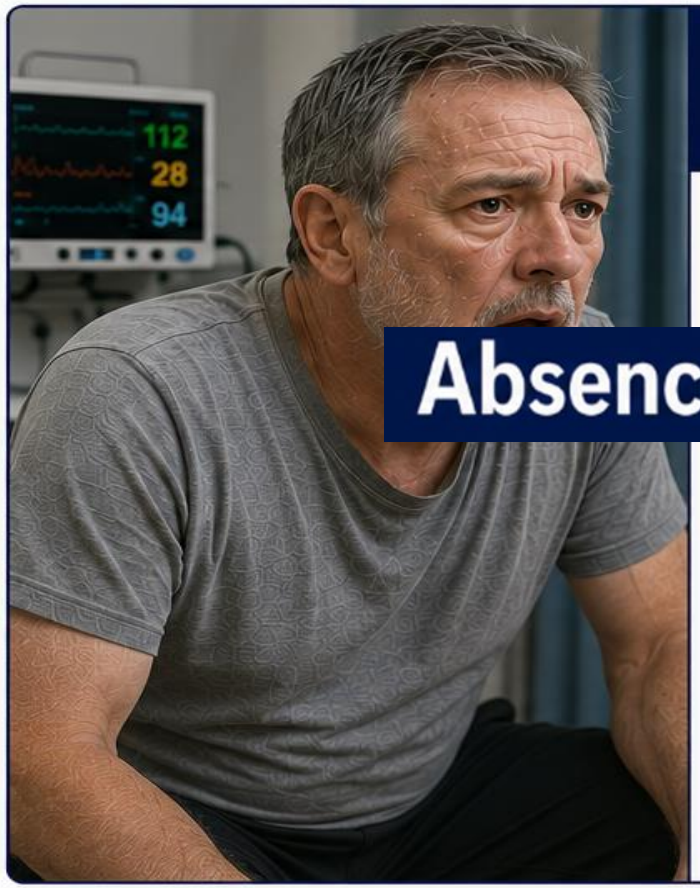
When Respiratory Failure… Doesn't Look Like Respiratory Failure



Traditional Respiratory Failure

- Tachypnea
- Labored breathing
- Marked dyspnea
- Easily recognized deterioration

Neuromuscular Respiratory Failure

- May appear comfortable
- Shallow respirations
- Fatigue or daytime sleepiness
- Subtle decline over months

Absence of distress does NOT equal stability.

NEUROMUSCULAR PATIENTS
FAIL VENTILATION BEFORE OXYGENATION

NORMAL RESPIRATORY SYSTEM

- Adequate respiratory muscle strength
- Effective alveolar ventilation
- Normal CO_2 elimination
- Normal sleep architecture

RESPIRATORY MUSCLE WEAKNESS

↓

HYPOVENTILATION

↓

HYPERCAPNIA

↓

SYMPTOMS

↓

RESPIRATORY FAILURE

NEUROMUSCULAR DISEASE

- Progressive respiratory muscle weakness
- Reduced tidal volume
- Alveolar hypoventilation
- CO_2 retention
- Sleep-disordered breathing

NORMAL SpO_2 DOES NOT EXCLUDE SIGNIFICANT HYPOVENTILATION.

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Supplemental Oxygen May Mask Ventilatory Failure

The patient may look better, but the problem may be getting worse.

BEFORE SUPPLEMENTAL OXYGEN

Physiologic response to hypoxemia triggers visible distress.

SpO_2	86%
Heart Rate	118
Respiratory Rate	32
Work of Breathing	Marked
Anxiety/Distress	High

These signs often prompt clinicians to administer oxygen.

SUPPLEMENTAL OXYGEN ADMINISTERED

AFTER SUPPLEMENTAL OXYGEN

Improved appearance may falsely reassure patients and clinicians.

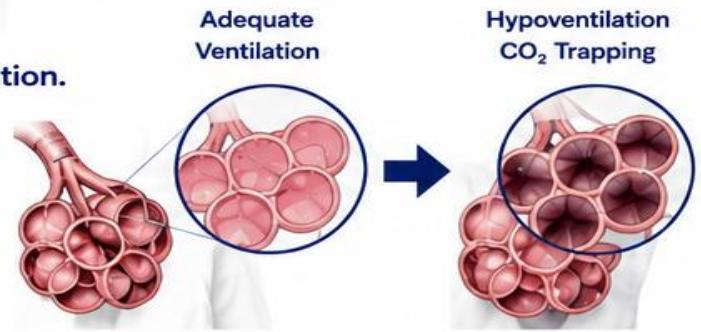
SpO_2	96%
Heart Rate	92
Respiratory Rate	20
Work of Breathing	Mild
Anxiety/Distress	Low

Visible distress decreases, but ventilation has not improved.

WHAT'S REALLY HAPPENING

Oxygen improves SpO_2, but does not improve ventilation.

- ⊗ Respiratory muscle weakness persists
- ⊗ Alveolar hypoventilation persists
- ⊗ CO_2 retention continues to worsen



CO_2 LEVELS CONTINUE TO RISE

$PaCO_2$	45 mmHg	----→	60 mmHg	----→ 80+ mmHg
	Early		Progressing	Decompensating

Rising CO_2 – Worsening Ventilatory Failure

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Clinical Clues to Respiratory Decline

Recognizing decline often requires looking beyond obvious respiratory distress.

Clinical Presentation

- Morning headaches
- Orthopnea
- Daytime sleepiness
- New daytime naps
- Frequent awakenings
- Shallow respirations

Objective Findings

- Elevated CO_2
- Elevated bicarbonate
- Declining FVC
- >15–20% drop in supine FVC
- Reduced inspiratory pressures
- Weak cough

Escalating Concern

- Increasing NIV dependence
- Increased secretion burden
- Inability to lie flat
- Difficulty speaking in full sentences
- Progressive daytime dyspnea
- Recurrent respiratory infections

Subtle symptoms and objective findings often identify respiratory decline before overt respiratory distress develops.

RECOGNIZING PROGRESSION:
OBJECTIVE MARKERS & WHEN TO ACT

Look for trends. Interpret in the context of symptoms, disease trajectory, and patient goals.

OBJECTIVE MARKER	CONCERNING FINDINGS	CLINICAL IMPLICATION
FVC (Forced Vital Capacity)	**< 50% predicted** or rapid decline	• **Evaluate for NIV initiation** *Earlier intervention improves outcomes*
SUPINE FVC (Supine Vital Capacity)	**> 15–20% decline** from sitting	• **Suggests diaphragmatic weakness** *Consider NIV evaluation*
MIP / SNIP (Inspiratory Muscle Strength)	**MIP < -60 cmH_2O** **or SNIP < 40 cmH_2O** (or progressive decline)	• **Significant inspiratory muscle weakness** *Consider ventilatory support*
CO_2 MONITORING (Gas Exchange)	**$PaCO_2$ or $TcCO_2$ > 45 mmHg** **or rising serum HCO_3^- > 27 mEq/L**	• **Evidence of chronic hypoventilation** *Initiate or escalate NIV*
SYMPTOMS (Clinical Indicators)	**Orthopnea, morning headaches, hypersomnolence, daytime fatigue, non-restorative sleep**	• **Evaluate for nocturnal hypoventilation and sleep-disordered breathing**
NIV UTILIZATION (Current Ventilatory Support)	**Increasing daytime use, persistent symptoms despite nocturnal NIV**	• **Evaluate for daytime ventilation strategies** *(mouthpiece ventilation, extended NIV support, or continuous support)*

DO NOT WAIT FOR OVERT RESPIRATORY FAILURE.
Symptoms and physiologic changes often precede acute decompensation.

KEY PRINCIPLE
No single value should drive intervention.
Look at the **trend**. Look at the **patient**.

FVC = Forced Vital Capacity | MIP = Maximal Inspiratory Pressure | SNIP = Sniff Nasal Inspiratory Pressure
$PaCO_2$ = Arterial CO_2 | $TcCO_2$ = Transcutaneous CO_2 | HCO_3^- = Bicarbonate

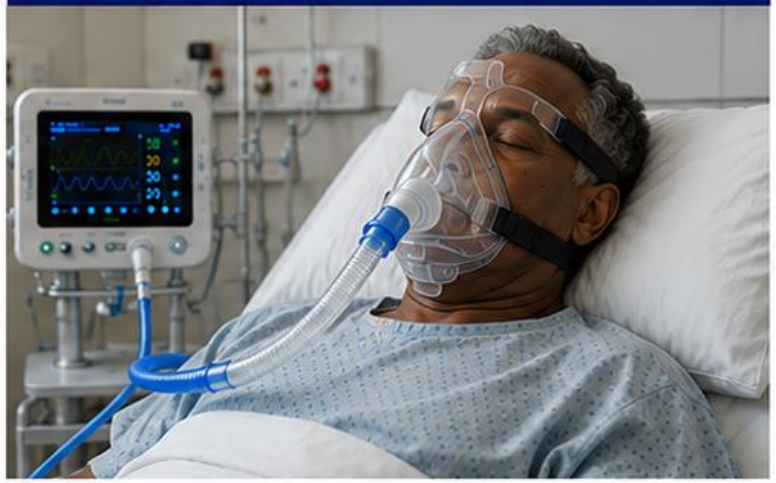
GOAL: EARLY RECOGNITION. PROACTIVE INTERVENTION. BETTER OUTCOMES.

Support that evolves with every breath.

Many patients are hospitalized for conditions such as pneumonia, secretion issues, or other complications before ventilatory failure occurs.

Recognizing hypoventilation early allows us to start NIV at the bedside—before failure.

NOCTURNAL OR HOSPITALIZATION NIV



Initiate early in hospital or at night

- Treat nocturnal hypoventilation
- Many patients first meet NIV during hospitalization (e.g., PNA, secretion issues)
- Start at bedside before failure occurs

DAYTIME MOUTHPIECE VENTILATION

Escalate support as daytime needs emerge

- Identify during an admission through assessment
- Mouthpiece ventilation provides support while preserving speech, mobility, eating, and independence
- Avoids the need for a mask all day

TRACHEOSTOMY VENTILATION

Consider when NIV no longer meets patient needs

- Persistent hypercapnia despite optimized NIV
- Inability to manage secretions / bulbar decline
- Inability to maintain adequate ventilation non-invasively
- Patient goals and preferences

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How Ventilatory Support Evolves in Neuromuscular Disease

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Optimizing NIV: Success Requires More Than Pressure

Ensure Mandatory Ventilation

- Backup respiratory rate is essential
- Spontaneous breathing alone may become insufficient
- Ensure adequate mandatory ventilation is delivered

Teaching Pearl: Weak muscles cannot sustain ventilation indefinitely.

Understand Your Modes

- Fully understand how the mode is biking (ST + backup rate)
- Recognize signs your patient is transitioning to VAPS
- Understand how your ventilator delivers pressure
- VAPS adjusts PS to achieve target ventilation

Teaching Pearl: Escalating ventilatory needs do not necessarily mean NIV has failed.

Comfort Settings Matter

- Trigger sensitivity
- Rise time
- Ti Min & Ti Max
- Cycling criteria
- Interface selection & humidification

Teaching Pearl: Comfort settings directly influence synchrony and effective ventilation delivery.

Hypercapnia ≠ Always More Pressure

- Assess for air leaks and optimize
- Look for adherence
- Evaluate ventilator mechanics
- Investigate infection
- Make appropriate adjustments
- Consider patient's thoracic and skeletal physiology

Teaching Pearl: Persistent hypercapnia should trigger troubleshooting—not automatic pressure escalation.

Before concluding NIV has failed, ensure therapy has truly been optimized.

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Case Study: Respiratory Failure Hiding in Plain Sight

Patient Presentation

- 36-year-old male with Duchenne Muscular Dystrophy
- Presented after being found down following a fall at home
- PMH: Restrictive lung disease, dysphagia, aspiration pneumonia, multiple falls, severe malnutrition

Hospital Course

- Initially conversational and clinically stable
- No respiratory evaluation performed until ~10 hours after admission
- At ~19:45, patient found to be:
  - Lethargic
  - In respiratory distress
  - Acutely altered
- **ABG/VBG obtained and patient was intubated for acute hypercarbic respiratory failure**

Initial VBG (at time of decompensation)

pH, Venous (T)	7.30	↓	(7.32 – 7.43)
pCO_2, Venous (T)	83	↑	(25 – 44 mmHg)
pO_2, Venous (T)	41		(25 – 44 mmHg)
HCO_3, Venous	40.8	↑	(22 – 29 mmol/L)
Base Excess, Venous	14.4	↑	(-2 – 3 mmol/L)
O_2 Sat, Venous	70		(%)

Audience Question: Looking back, what clues suggest this patient was experiencing chronic hypoventilation before admission?

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Case Study: Optimization Changed the Trajectory

1. Admission

Found down after fall

Initially stable

No respiratory eval

2. Acute Decompensation

~10 hrs after admission

Lethargic, in distress

VBG: pCO_2 83

→ Intubated

3. Post-Intubation

Treated for aspiration pneumonia

Supportive care

4. Extubation

Extubated on HD #2

Transitioned to NIV (settings similar to home device)

5. NIV Escalation

Persistent hypercapnia

→ Increased EPAP and PS

6. Worsening Gas

Despite higher pressures:

pH 7.17

pCO_2 102

HCO_3 31

7. RT Consultation

Saw-tooth waveform on Trilogy EV300

Severe thoracic restriction

Pressure-limited

8. Optimization

Strategy redesigned (see below)

Improved alveolar ventilation

9. Discharge

Home on optimized NIV settings

Outpatient follow-up

Initial Decompensation (Pre-Intubation)

VBG

pH	7.30
pCO_2	83
HCO_3	40.8

→

Post-Extubation NIV (Similar to Home Settings)

VBG

pH	7.22
pCO_2	90
HCO_3	58

→

After NIV Escalation (↑ EPAP & PS)

VBG

pH	7.17
pCO_2	102
HCO_3	31

→

Optimization Strategy

- Reduced EPAP to increase pressure window
- Adjusted PS to deliver effective VT without pressure limiting
- Optimized rise time, Ti, and cycle for patient comfort
- Improved synchrony and reduced saw-tooth pattern
- Focused on lower lung (basal) ventilation

→

Discharge (Optimized NIV)

VBG

pH	7.42
pCO_2	51
HCO_3	31

→

Outpatient Follow-Up

VBG

pH	7.41
pCO_2	38
HCO_3	25

Key Outcome

- ✓ Successfully extubated
- ✓ Hypercapnia resolved
- ✓ Discharged on optimized NIV
- ✓ Outpatient respiratory follow-up
- ✓ Ultimately achieved normal gas exchange

Teaching Pearl: *NIV failure is frequently optimization failure. Understanding patient physiology and ventilator mechanics can dramatically alter outcomes.*

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CLINICAL REASONING CHALLENGE

The VBG continued to worsen despite escalating NIV settings (IPAP 20 → 25 cmH_2O; EPAP 3 → 5 cmH_2O).

What was the most likely physiologic consequence of these pressure increases?

A Central apnea developed, requiring a higher backup rate

B Increased pressures contributed to patient-ventilator desynchrony and worsening work of breathing

C Pressure limitation reduced effective pressure support and resulted in inadequate tidal volume delivery

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Thank You

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Special thanks to:

The California Society for Respiratory Care (CSRC)
for their commitment to advancing respiratory care education and professional development.

Scripps Health
for providing the opportunity to share this work and foster ongoing learning in neuromuscular respiratory care.

The UC San Diego Health Leadership Team
for their continued support and collaboration in:

- Developing and implementing our neuromuscular respiratory care pathway
- Establishing and advancing the Ambulatory Respiratory Therapist Treatment Team
- Expanding bedside clinician knowledge through education focused on neuromuscular disease and chronic respiratory failure

Together, through collaboration, education, and innovation,
we can continue to improve outcomes for patients living with neuromuscular disease.

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