Managing Challenges and Maximizing Success in Mechanical Ventilation

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Disclosures

- Consultant, Scientific Advisory Board, or Lecturer
  - Hoffman-La Roche
  - Scholar Rock
  - Biogen
  - Sanofi
  - PTC Therapeutics
  - Catalyst Pharmaceuticals
  - RegenX Bio

Outline

- What is hypoventilation
- What is the impact of hypoventilation and management
- When to look for hypoventilation
- How to evaluate for hypoventilation
- How to treat hypoventilation
- Managing challenges
- Maximizing success

What is Hypoventilation

What is Hypoventilation?

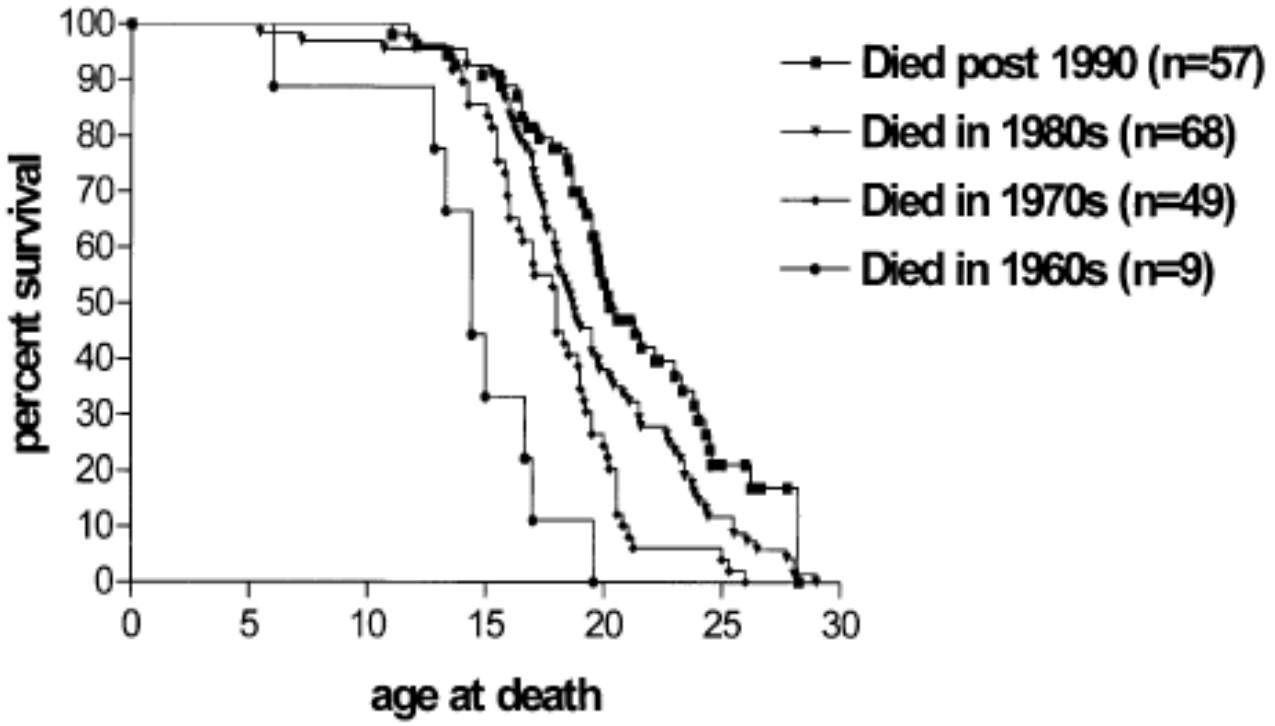
- Underventilation due to
  - Inadequate minute ventilation
    - Tidal Volume (size of breath)
    - Respiratory Rate
  - Inadequate drive to breathe
    - Pharmacologic
    - Acute
    - Chronic
- Outcome
  - Hypercarbia
  - Hypoxemia

Impact of Hypoventilation

Impact of Hypoventilation

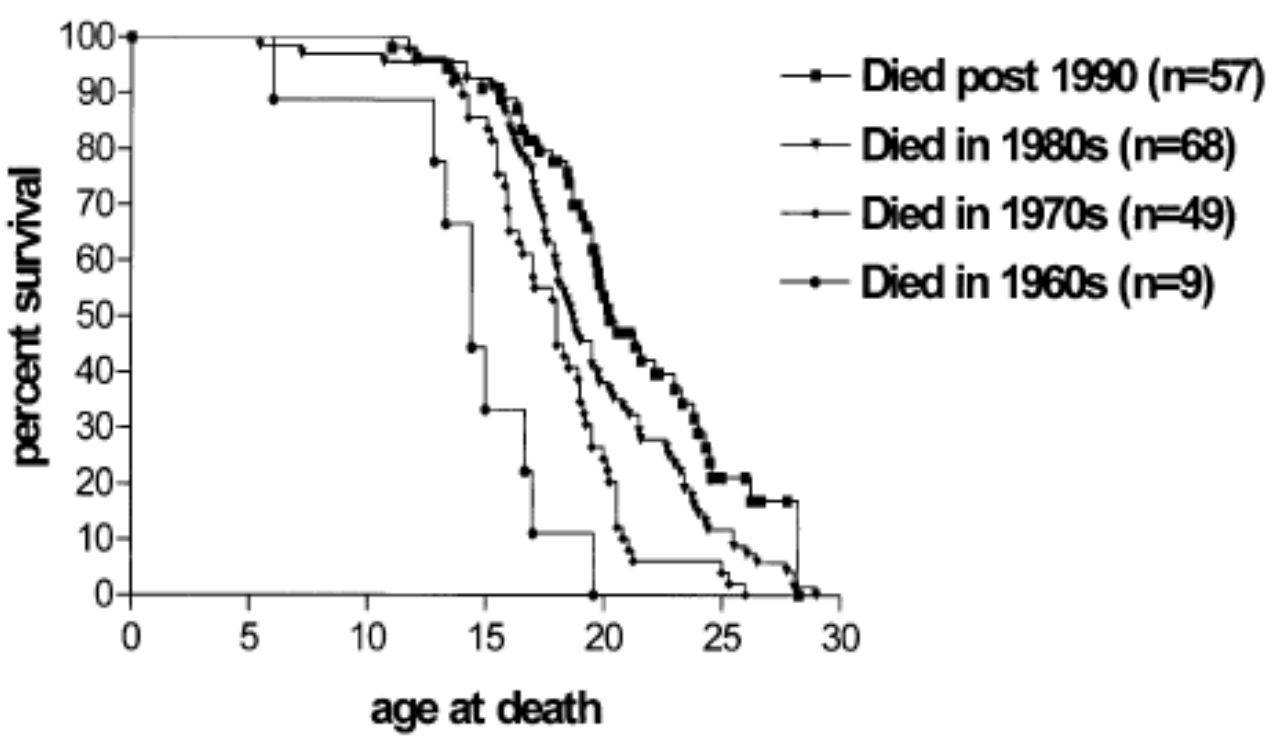
- Response
  - Hypercarbia
    - Increased respiratory rate and work of breathing
    - Taking calories from other needs
    - Renal compensation
  - Hypoxemia
    - Pulmonary hypoxic vasoconstriction
    - Increased pulmonary blood pressure
    - Increased right heart strain
- Decreased energy
- Weight loss / failure to thrive

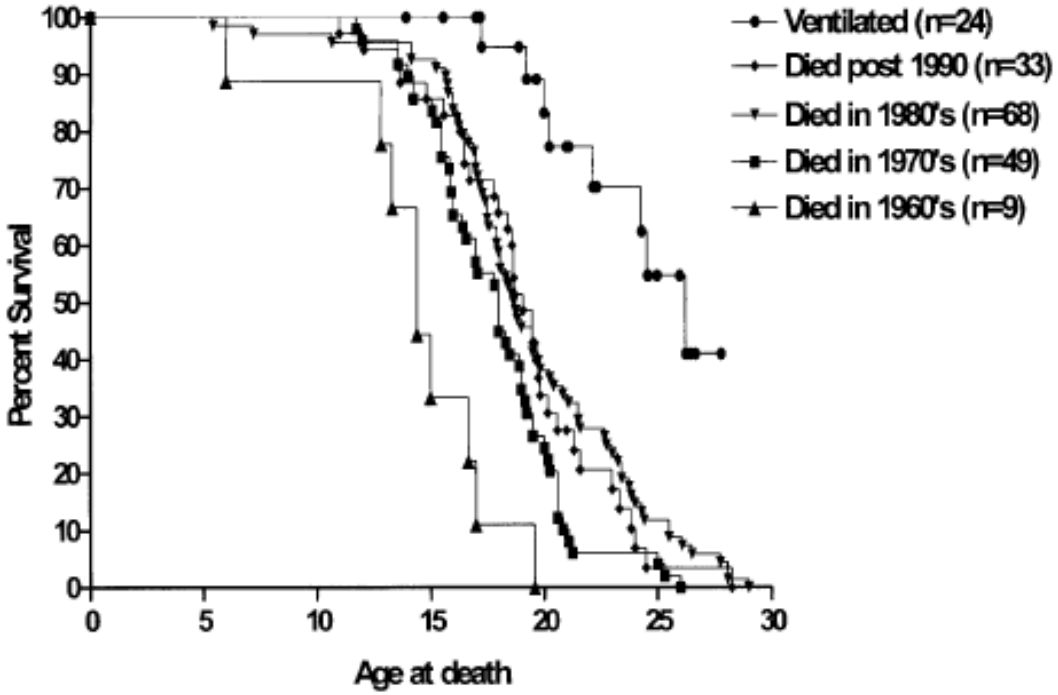
Impact of Managing Hypoventilation



Eagle, *et.al. Neuromuscular Disorders* 2002

Impact of Managing Hypoventilation





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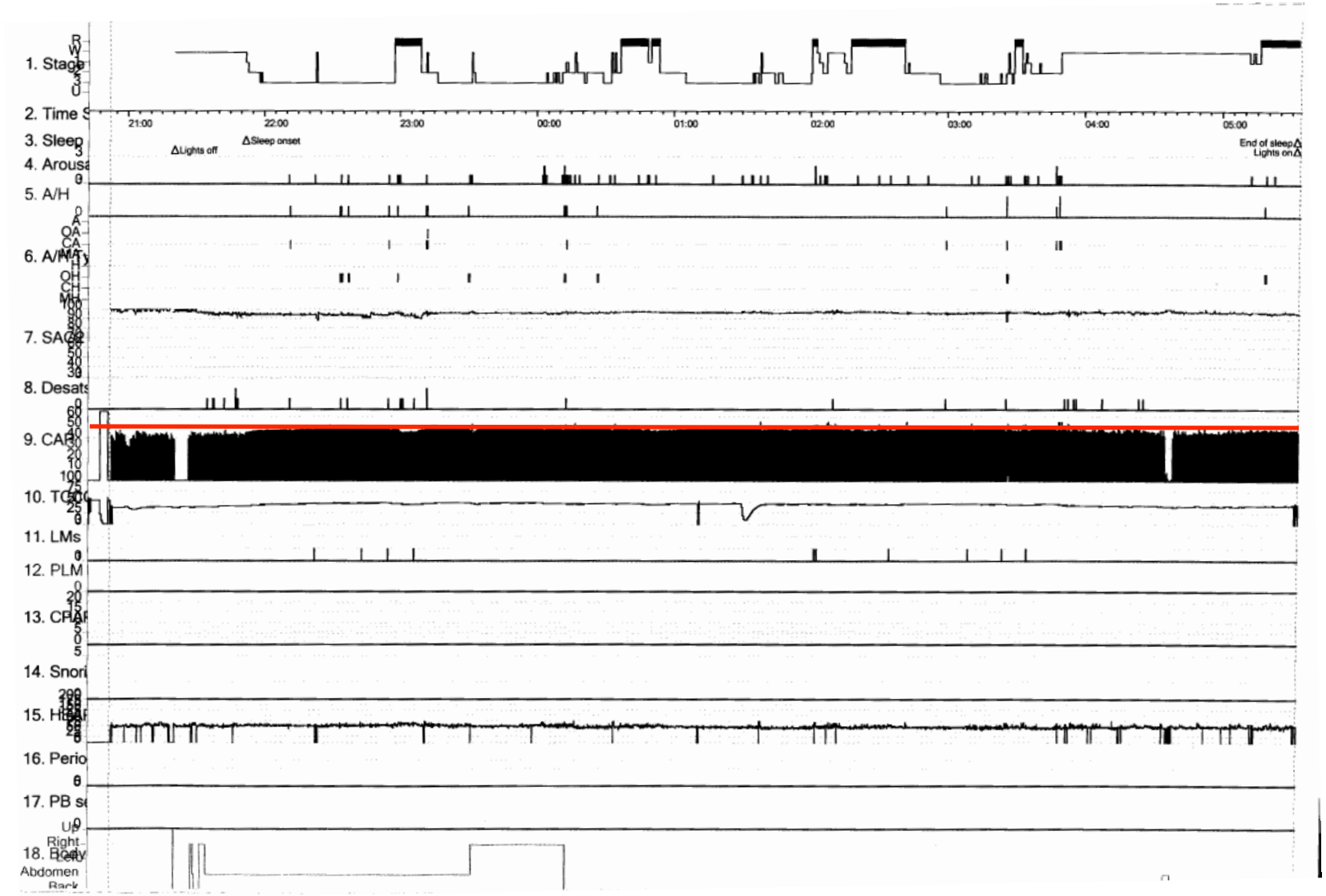
How to Evaluate for Hypoventilation

How to Look for Hypoventilation



How to Look for Hypoventilation

ETCO2 Intervals	>60	50-60	45-50	35-45	<35
Sleep Time (%)	0.0	17.2	81.1	1.7	0.0

ETCO2 & TCCO2 Summary

	Maximum (Torr)		Mean (Torr)	
	ETCO2	TCCO2	ETCO2	TCCO2
Awake	52	50	44	44
REM	50	49	47	47
NREM	53	54	49	48
Total Sleep	53	54	48	48

Oxygenation Summary

SpO2 %	>= 95%	< 95%	< 92%	< 90%	< 88%	< 85%	< 80%	< 70%	< 60%
Sleep Time(minutes)	300.8	60.1	3.6	0.2	0.0	0.0	0.0	0.0	0.0
Sleep Time (%)	82.6	16.5	1.0	0.1	0.0	0.0	0.0	0.0	0.0

	SpO2 Min. (%)	SpO2 Max (%)	SpO2 Mean (%)
REM	92	99	97
NREM	88	99	97
Total Sleep	88	99	97

Respiratory Event Summary

			Obstructive			OAHI	Central		CAHI	Total
			Apnea	Hypop	Mixed		Apnea	Hypop		
Total		Count	1	10	0	11	7	0	7	18
		Index	0.2	1.6	0.0	**1.8**	1.2	0.0	1.2	3.0
REM		Count	0	2	0	2	0	0	0	2
		Index	0.0	1.7	0.0	1.7	0.0	0.0	0.0	1.7
Position (all stages)	Supine	Count	0	3	0	3	6	0	6	9
		Index	0.0	0.8	0.0	0.8	1.6	0.0	1.6	2.4
	Prone	Count	1	5	0	6	1	0	1	7
		Index	0.6	3.0	0.0	3.7	0.6	0.0	0.6	4.3
	Side	Count	0	2	0	2	0	0	0	2
		Index	0.0	2.9	0.0	2.9	0.0	0.0	0.0	2.9

How to Look for Hypoventilation

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ETCO2 & TCO2 Summary

	Maximum (Torr)		Mean (Torr)	
	ETCO2	TCO2	ETCO2	TCCO2
Awake	52	5	44	44
REM	50	4	47	47
NREM	53	5	49	48
Total Sleep	53	5	48	48

Oxygenation Summary

SpO2 %	>= 95%	< 95%	< 92%	< 90%	< 88%	< 85%	< 80%	< 70%	< 60%
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Nocturnal Pulse Oximetry is Not a Viable Alternative

Hypoventilation Assessment - Caution

- Significant vs. Non-Significant hypercarbia
  - Is $EtCO_2 > 50$ for < 20% sleep time Normal?
- Patients are typically studied when well
  - What about during illness
- Access to formal Sleep Study
  - Most testing not for hypoventilation
    - No capnography needed for OSA assessment
  - Small number of centers capable of doing a pediatric sleep study
  - Timeliness?
    - Delay in getting in for a diagnostic study

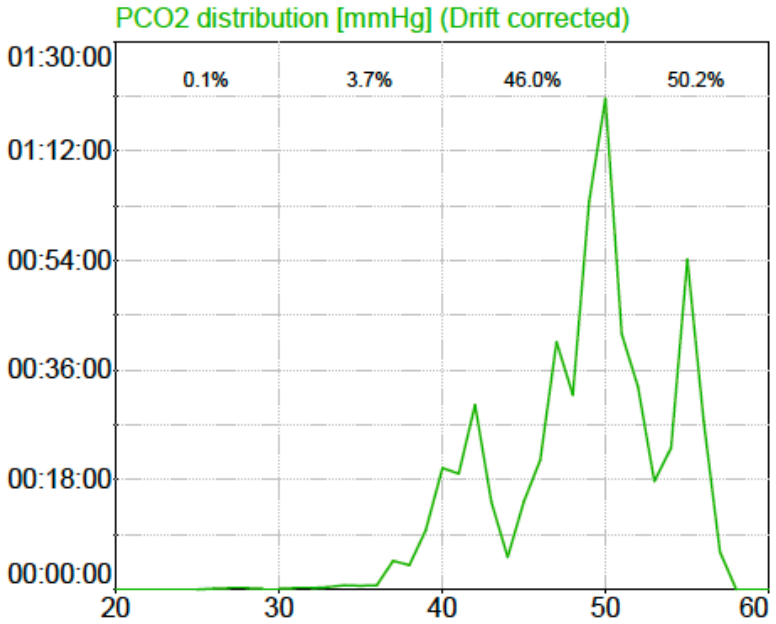
Hypoventilation Assessment - Caution

- OSA vs Hypoventilation
  - Hypopnea
    - 50% decrease in nasal air flow
    - Thoracoabdominal asynchrony
    - Mild oxyhemoglobin desaturation (5%)
- OSA – Obstruction as the cause
  - Elevated Apnea-Hypopnea Index (AHI) with both hypopneas AND apneas
  - Hypopneas – low flow due to flow limitation/obstruction
- Hypoventilation – Underbreathing as the cause
  - Elevated AHI with no or very few apneas
  - Hypopneas – low flow due to effort / weakness AND No flow limitation
- Poor guidance in the Sleep Medicine Guidelines

Hypoventilation Simplified

- Novel approaches for assessment
  - Inpatient nocturnal capnography
    - EtCO2
    - TcCO2
    - Uncomfortable bed
  - Home nocturnal capnography
    - TcCO2
    - Comfortable bed
    - Real life assessment
    - Relatively easily done

Home Capnography



		PCO2 Range [mmHg]	Duration	%-Duration	%-Integrated
Minimum (occur.)	26.3 mmHg (06:16:29)	90.0 - 200.0	00:00:00	0.0	100.0
Maximum (occur.)	56.7 mmHg (02:36:37)	85.0 - 89.9	00:00:00	0.0	100.0
Mean	48.7 mmHg	80.0 - 84.9	00:00:00	0.0	100.0
Median	49.5 mmHg	75.0 - 79.9	00:00:00	0.0	100.0
Baseline (occur.)	49.3 mmHg (21:01:13)	70.0 - 74.9	00:00:00	0.0	100.0
Max. above baseline (occur.)	7.4 mmHg (02:36:37)	65.0 - 69.9	00:00:00	0.0	100.0
		60.0 - 64.9	00:00:00	0.0	100.0
Time >50.0 mmHg	03:46:42	55.0 - 59.9	00:55:50	9.8	100.0
Time > 50.0 mmHg [%]	40	50.0 - 54.9	02:58:49	31.5	90.2
Events > 50.0 mmHg, duration > 5 min.	7	45.0 - 49.9	03:36:08	38.1	58.7
		40.0 - 44.9	01:22:58	14.6	20.6
		35.0 - 39.9	00:31:28	5.5	6.0

Hypoventilation – A New Approach

- Delphi Process
  - 3 rounds
  - Literature review to identify criteria for consideration
  - 15 pediatric pulmonologists with expertise in neuromuscular disease
  - 10 board certified in sleep medicine

Respiratory Insufficiency in Neuromuscular Disease (RIND): A Delphi Study to Establish Consensus Criteria to Define and Diagnose Hypoventilation in Pediatric Neuromuscular Disease

Oscar Henry Mayer[a,*], Reshma Amin[b], Hemant Sawnani[c], Richard Shell[d], Sherri Lynne Katz[e] and the RIND Study Group[1]

Mayer, *et. al. J Neuromusc Dis.* 2023

Hypoventilation – A New Approach

- Normal
- Hypoventilation
  - CO2:
    - > 45 mmHg for 25% of sleep;
    - > 50 mmHg for >2% or 5 minutes continuously;
    - 10 mmHg above baseline
  - SpO2:
    - Mean < 94%
    - < 90% for >2% or 5 minutes continuously

Hypoventilation – A Newer Approach

- Normal
- *Transitional*
  - *Normal TcCO2/EtCO2*
  - *Tachypnea (25% above awake baseline)*
  - *Thoracoabdominal asynchrony / increased WOB*
- Hypoventilation
  - CO2:
    - > 45 mmHg for 25% of sleep;
    - > 50 mmHg for >2% or 5 minutes continuously;
    - 10 mmHg above baseline
  - SpO2:
    - Mean < 94%
    - < 90% for >2% or 5 minutes continuously

How We Can Use This Approach

- Objective definition of hypoventilation / respiratory failure
  - Not “Started BlPAP”
    - Clinical decision
    - Variable based on clinical practice
  - Can be used as a clear measure of progression / improvement
    - Identity absence of hypoventilation
- Provide a clinical spectrum / progression
- Can allow easier assessment and outside of the Sleep Lab

How to Treat Hypoventilation

Mechanical Ventilation

- Interface
  - Nasal
  - Oral
  - Oronasal
  - Tracheostomy
  - Full body
- Ventilator
  - Type
    - Intermittent
    - Continuous
- Mode
  - Pressure (Positive or Negative)
  - Volume

Nasal Masks

Canula Interfaces

Other Interfaces

Face Masks

Chin Strap

Mouthpiece Ventilation

Mouthpiece Ventilation

Tracheostomy Tube

Nocturnal Ventilation

Continuous Ventilation

Initiating Non-Invasive Ventilation

- Traditional – sleep lab
  - Access is difficult
- Alternative – inpatient
  - Expensive and capacity issues
- New Alternative – Office initiation / Home Confirmation
  - TcCO2 in office and at home

Utility of a Tracheostomy Tube

- Benefits
  - Most stable interface
  - Full ventilation during activities
  - Nothing on the face
- Challenges
  - Narrows effective airway
  - Need for surveillance by skilled caregivers
    - Emergent tracheostomy tube change

Need for a Tracheostomy Tube

- Absolute
  - Unstable upper airway
  - Unable to maintain a non-invasive interface
  - Unable to deliver pressures needed for ventilation non-invasively
- Relative
  - Prolonged duration of ventilation
    - Unable/unwilling to use Nasal/Mouthpiece ventilation
  - Discomfort using / intolerance of nasal or oral interface

Need for a Tracheostomy Tube

- Contraindication
  - Access to the neck
    - Short neck
    - Spinal deformity – cervical kyphosis (chin to the chest)
  - Unable to provide proper surveillance / safe environment
    - Trained caregivers (family)
    - Nursing
- Ultimate goal
  - Quality of life for the patient – personal decision

Managing Challenges

Challenges

- Interface
  - Mask size on face
    - Impact vision
  - Placement
    - Discomfort
    - Skin irritation
    - Skin injury
  - Proper seal
    - Air leak around mask contact on face
    - Poor patient / ventilator synchrony
  - Important to regularly assess fit and skin integrity

Challenges

- Ventilator
  - Triggering of breaths
    - Inspiration – getting a breath when you want
    - Exhalation – making the breath stop when you want
  - Size of breaths
    - Approximate normal tidal volume
    - Have the air go in at the right speed
      - Sensation of getting the breath that is needed
  - Accommodate for variable needs
    - Awake vs sleep
    - Well vs ill
  - Assess ventilation success
    - Ventilator data download
    - Capnography

Maximizing Success

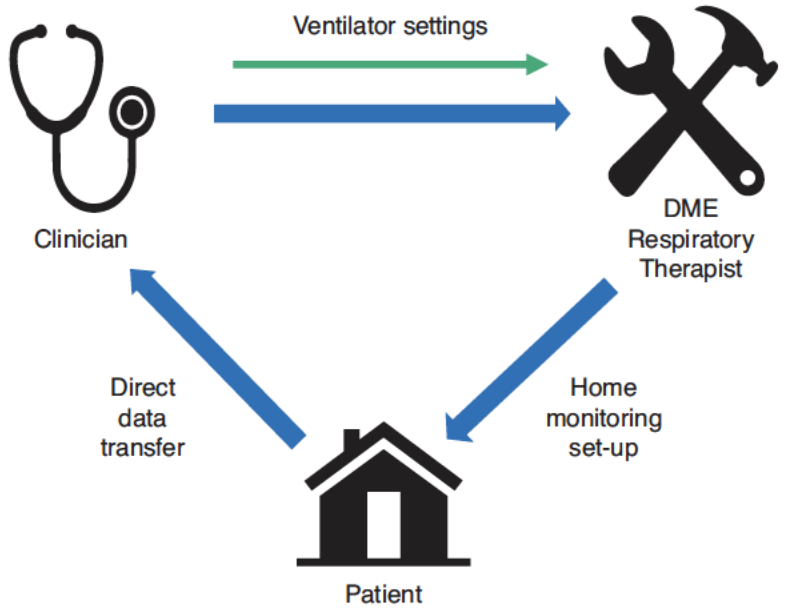
Maximizing Success

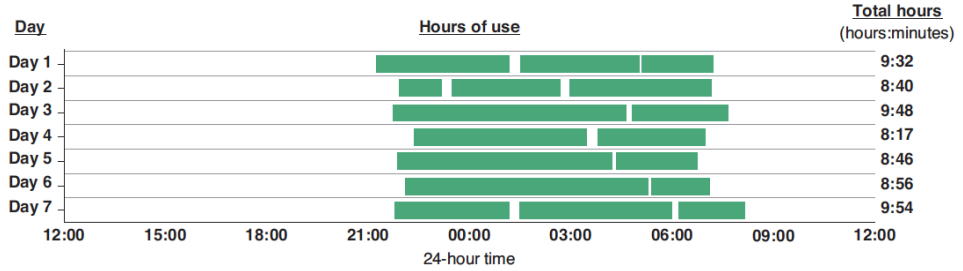
- Routine surveillance
  - In person – Physician Office
    - Patient-ventilator synchrony
    - Assess stability and energy
  - Ventilator data – can be extracted remotely if modem is installed
    - Attaining proper pressures
      - Leak in circuit (mask)
    - Usage
      - Days
      - Duration per day
    - Correlate to TcCO2 trending

Maximizing Success

- Routine surveillance
  - Gas exchange
    - Capnography
      - Serum HCO_3^-
      - $EtCO_2$
      - $TcCO_2$
    - Oximetry
  - Nocturnal ventilation
    - Capnography – $TcCO_2$
    - Sleep study

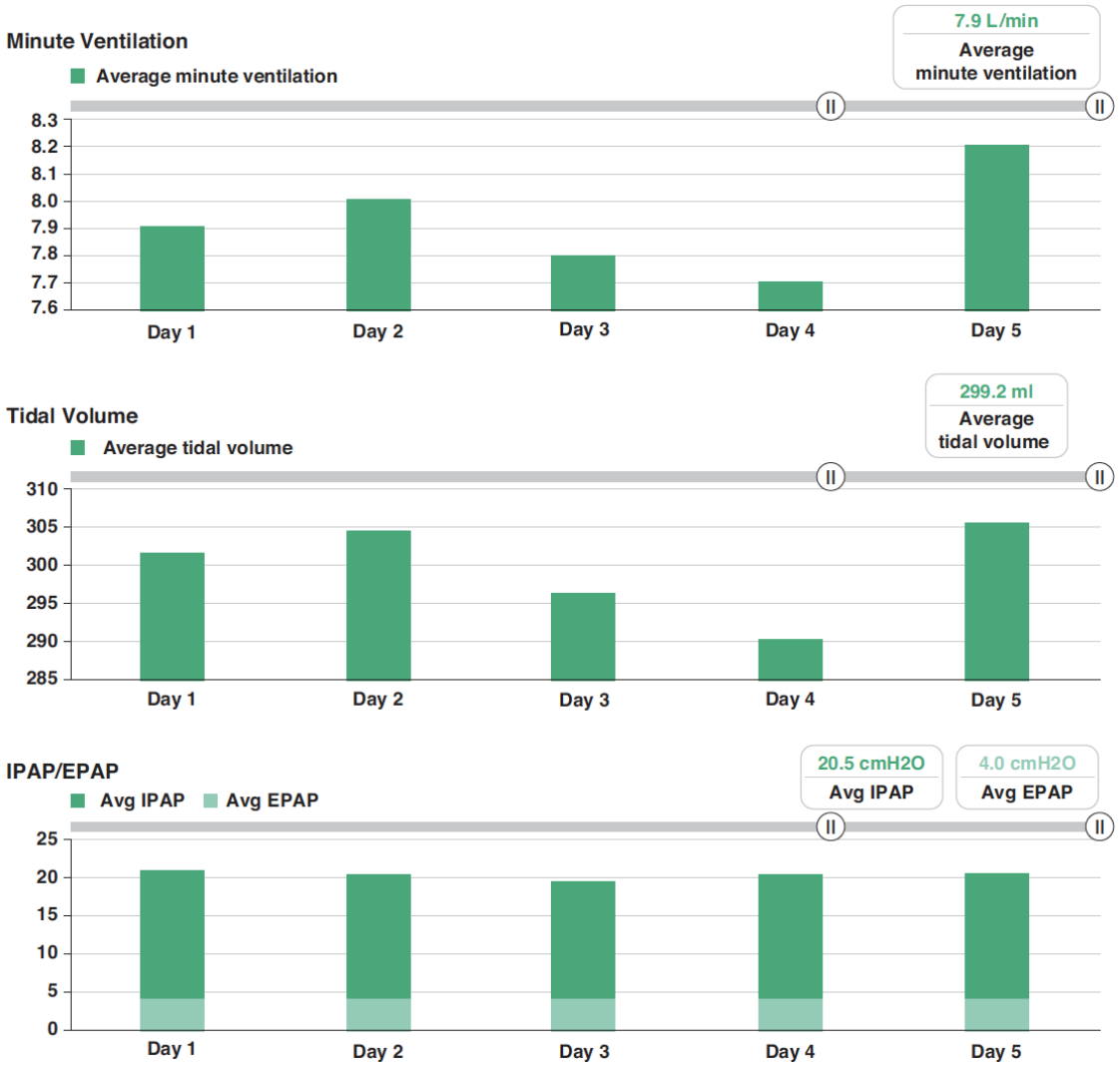
Outpatient Follow-Up of NIV





Ackrivo, *et. al. Annals ATS.* 2021

Outpatient Follow-Up of NIV



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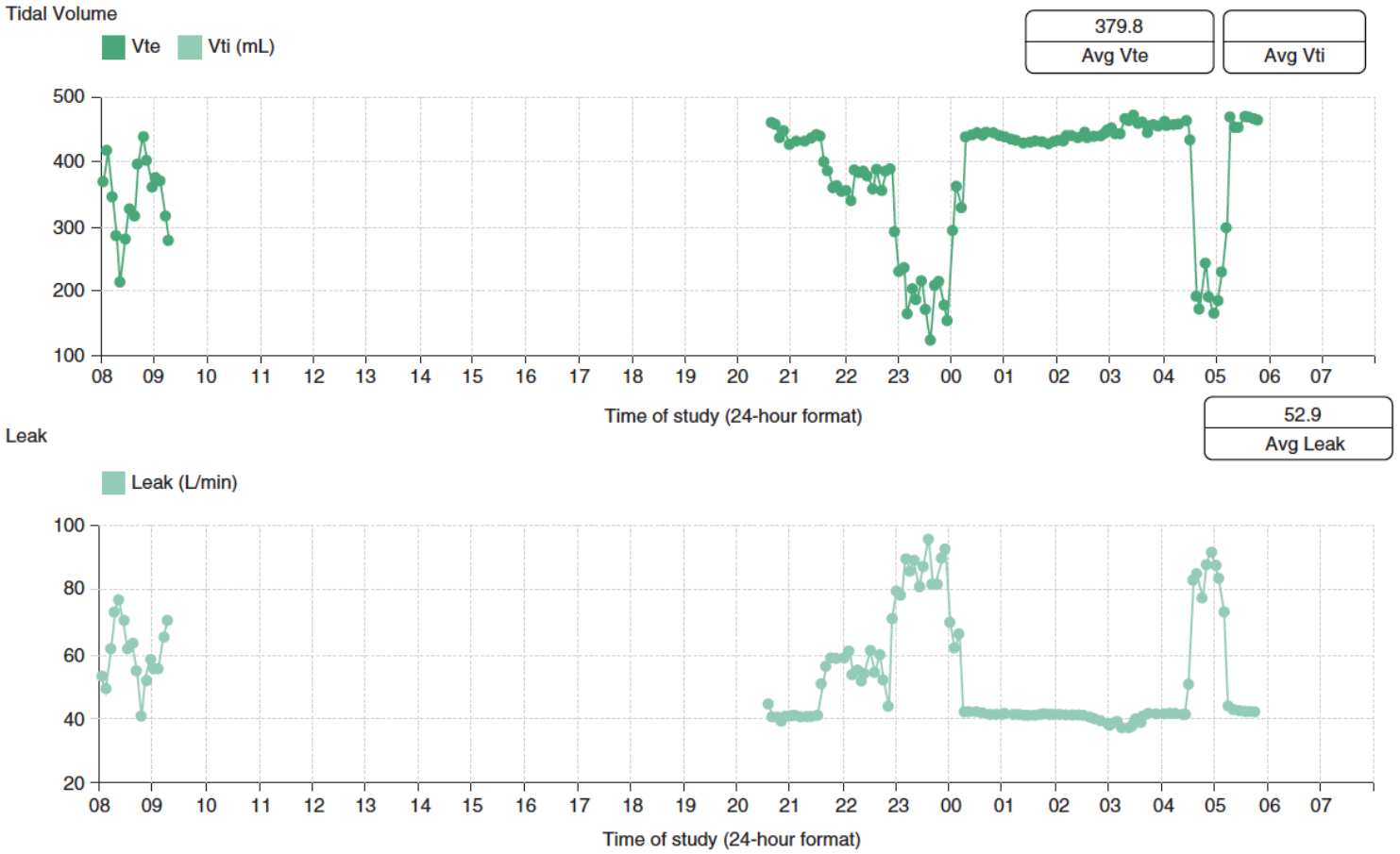

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Conclusion

- Treating hypoventilation is critical in giving patients the chance to reach their full potential
- There are many interfaces for and modes of ventilation and no rules on use and implementation
- Being flexible in implementing ventilatory support is critical in meeting a patient’s needs and maximizing success