Medical Service Company

20th ANNUAL JEAN S. MARX Memorial Education Forum

Care of Patients with Chronic Neuromuscular Respiratory Weakness

Beaumont

Nader Mina, MD

Medical Director; Sleep Evaluation Services

Division of Pulmonary, Critical Care, and Sleep Medicine, Beaumont, Royal Oak

Assistant Professor, Department of Internal Medicine

Oakland University William Beaumont School of Medicine

Disclosures

- None

Objectives

- Review of respiratory neuroanatomy
- General respiratory issues in neuromuscular disease (NMD)
- Sleep and neuromuscular disease
- Specific Diseases
- Respiratory evaluation of patients with NMD
- Management

Objectives

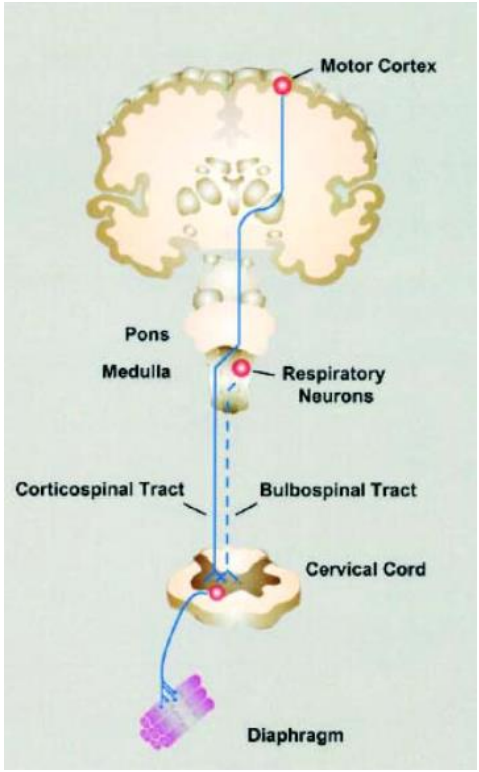
Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

- **Review of respiratory neuroanatomy**
- General respiratory issues in neuromuscular disease (NMD)
- Sleep and neuromuscular disease
- Specific Diseases
- Respiratory evaluation of patients with NMD
- Management

Respiratory Neuroanatomy

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

- **CNS**
  - -Chemoreceptors, medullary drive centers, spinal cord
- **Peripheral Nervous System**
  - -Nerve roots, peripheral nerves
- **Peripheral Receptors**
  - -Chemoreceptors, muscle receptors



Respiratory Neuroanatomy

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

Muscles of Inspiration

Core Muscles

- External intercostals
  (contracts to elevate ribs)
- Diaphragm
  (contracts to expand thoracic cavity)

Accessory Muscles

- Sternocleidomastoid
  (contracts to elevate sternum)
- Pectoralis minor
  (contracts to pull ribs outwards)

Muscles of Expiration

Core Muscles

- Internal intercostals
  (contracts to pull ribs down)
- Diaphragm
  (relaxes to reduce thoracic cavity)

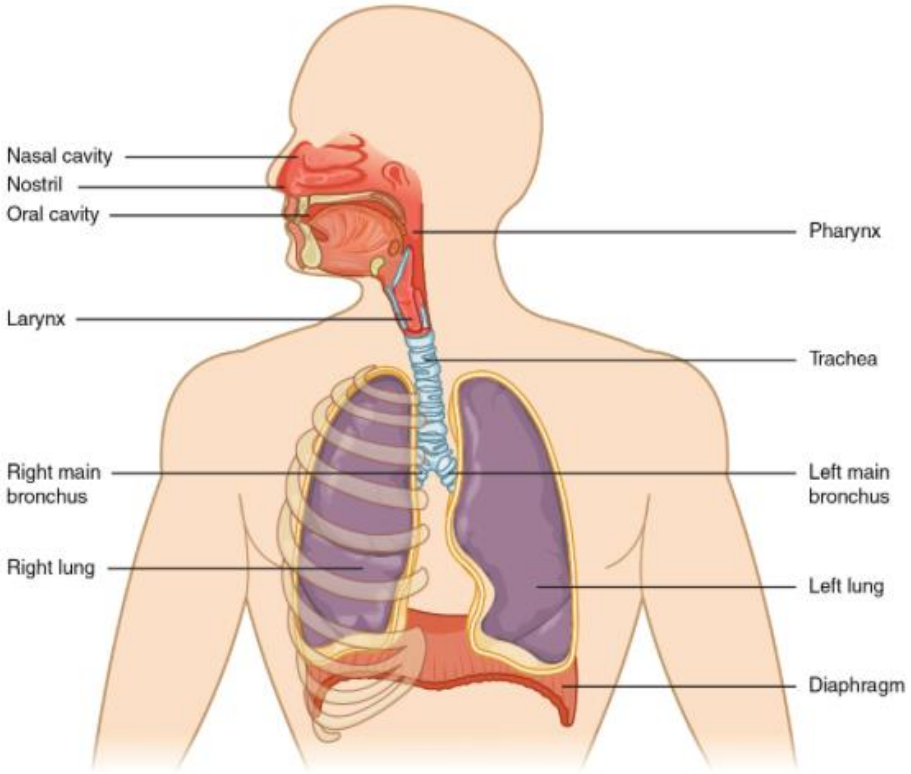
Accessory Muscles

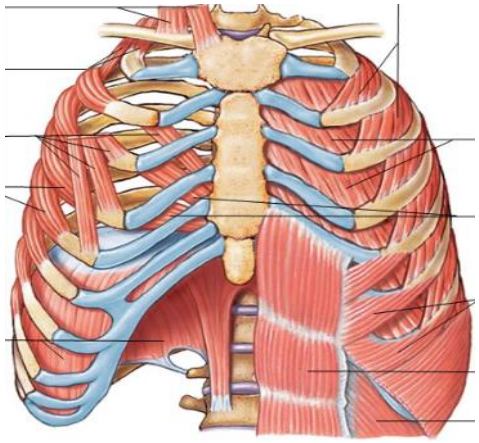
- Abdominals
  (contracts to compress abdomen)
- Quadratus lumborum
  (contracts to pull ribs down)

Respiratory Neuroanatomy

Medical Service Company

20th ANNUAL JEAN S. MARX Memorial Education Forum





Objectives

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

- Review of respiratory neuroanatomy
- **General respiratory issues in neuromuscular disease (NMD)**
- Sleep and neuromuscular disease
- Specific Diseases
- Respiratory evaluation of patients with NMD
- Management

Respiratory System and NMD

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

Weakness of respiratory muscles (inspiratory, expiratory, and upper airway)

Inadequate Ventilation

Nocturnal Hypoventilation

Bulbar Dysfunction

Ineffective Cough

Respiratory System and NMD

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

Inadequate Ventilation

- Weakness of inspiratory muscles
- Decrease in tidal Volume
- Increase to respiratory rate (inadequate compensation)
- Rise in PaCO2
- Hypoxemia (Due to hypoventilation and atelectasis)
- Symptoms and signs:
  - Dyspnea, orthopnea
  - Rapid shallow breathing
  - Accessory muscle use

Respiratory System and NMD

Bulbar Dysfunction

- Impairment of upper airway muscles
- Symptoms and signs:
  - Dysphagia
  - Dysarthria
  - Weak mastication
  - Facial weakness
  - Abnormal secretions clearance
  - Nasal speech
  - Protruding tongue

Respiratory System and NMD

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

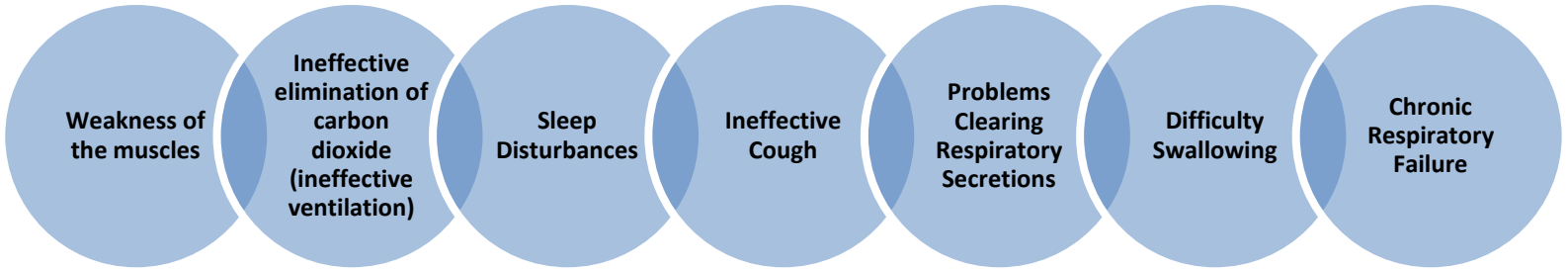
Ineffective Cough

- Weakness of upper airway muscles, inspiratory, and expiratory muscles

- Mechanism of cough:
  - Inspiratory phase => Large rapid tidal volume inspiration
  - Compressive phase => Glottic Closure (by addutor muscles) => increase in positive intrathocic pressure
  - Expiratory phase => Glottic opening => high peak expiratory flow

Respiratory System and NMD

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum



Objectives

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

- Review of respiratory neuroanatomy
- General respiratory issues in neuromuscular disease (NMD)
- **Sleep and neuromuscular disease**
- Specific Diseases
- Respiratory evaluation of patients with NMD
- Management

Sleep and NMD

Physiologic changes in Respiratory System during Sleep

Control of Breathing:

Wake: Behavioral (Cortical Centers)

Metabolic

Sleep: Only Metabolic (PH, PaO_2, $PaCO_2$)

Reduce ventilatory responses

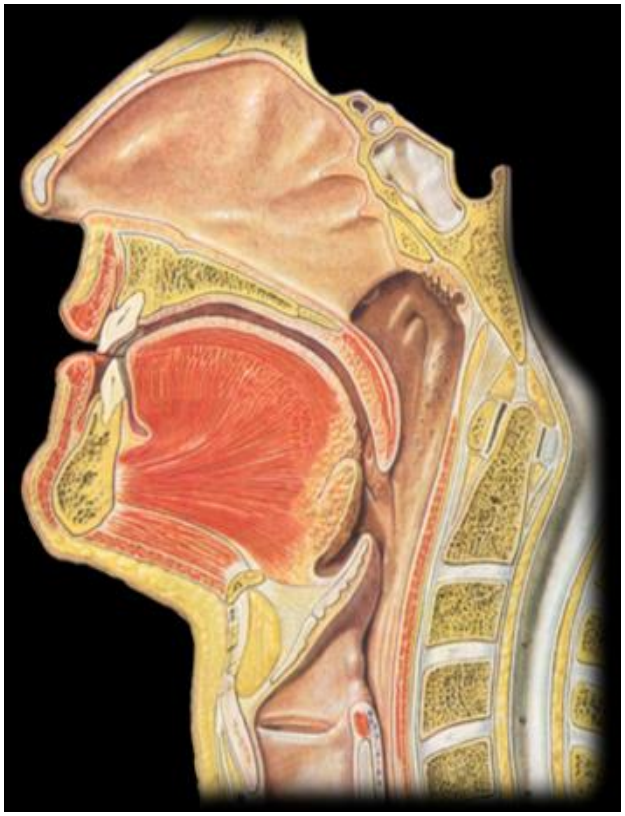
Sleep and NMD

Physiologic changes in Respiratory System during Sleep

Airways and Respiratory Muscles:

↓↓ UA dilator muscle tone => Increase Collapsibility

↓↓Activity of accessory muscles of respiration
=> Decrease Minute Ventilation



Mohsenin, Semin Resp Crit Care Med 2005;26:109

Sleep and NMD

Summary of Important Physiologic Changes

- Decreas inute
- Decrease in
- Dec
- Increase
- FRC ss intercostal muscle activit

Respiratory System is Vulnerable During Sleep

Mohsenin, Semin Resp Crit Care Med 2005;26:109

Sleep and NMD

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

Changes in patients with NMD

- Weakness of respiratory and chest wall muscles
- Weakness of upper airway muscles => increase airway resistance
- REM- related hypotonia and atonia of all muscles except diaphragm => diaphragmatic overload => Nocturnal Hypoventilation (usually before daytime hypoventilation)
- Kyphoscoliosis => restrictive disease
- Reduced responsiveness of chemoreceptors due to altered afferent input from skeletal muscle spindles

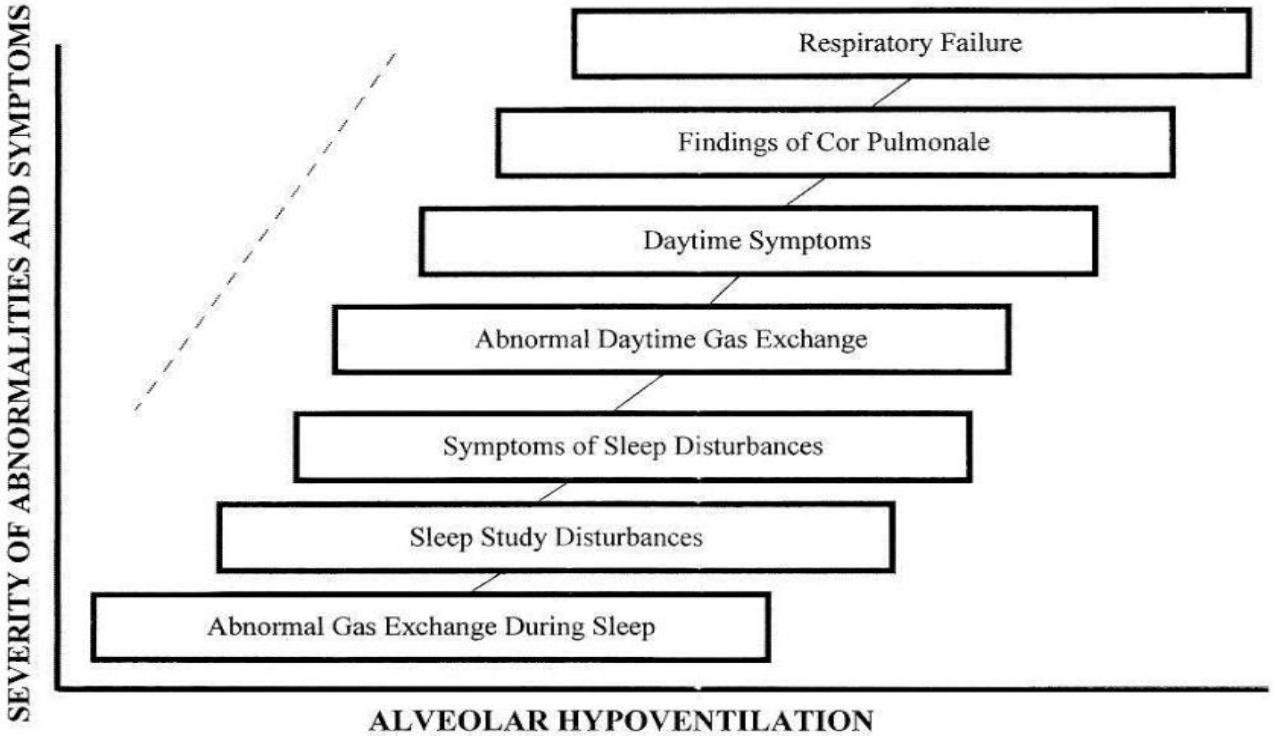
Sleep and NMD

Symptoms and Signs:

- Poor concentration
- Drop in school or work performance
- Nocturnal awakenings
- Morning headaches and fatigue
- Hypersomnolence
- Impaired cognition

Sleep and NMD

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum



Chest, Volume 116 (2)-August 1999

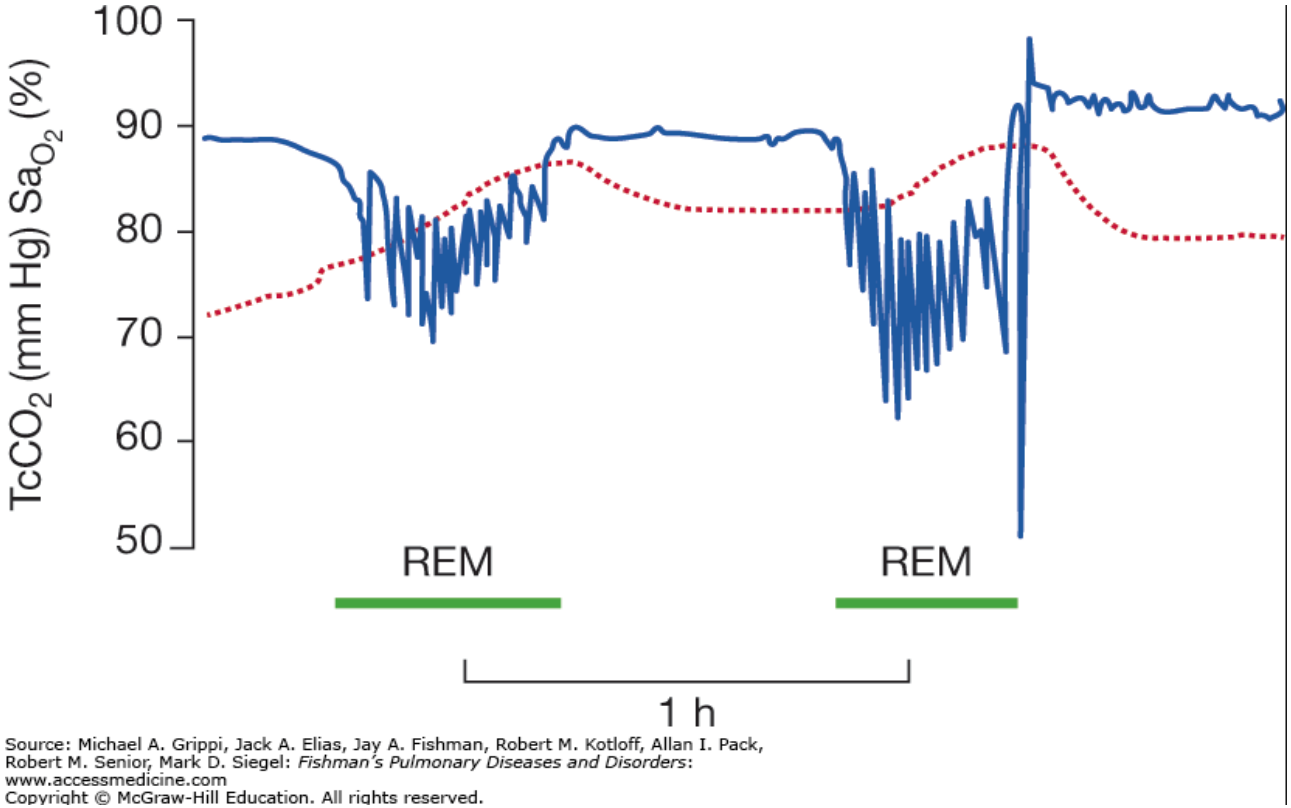
Sleep and NMD

Polysomnographic Findings

- Reduced lung volumes when supine => severe desaturations especially in REM sleep
- Hypoventilation with marked CO2 elevation
- Pseudocentral apneas
- Obstructive apnea due to upper airway weakness
- Decreased REM sleep

Sleep and NMD

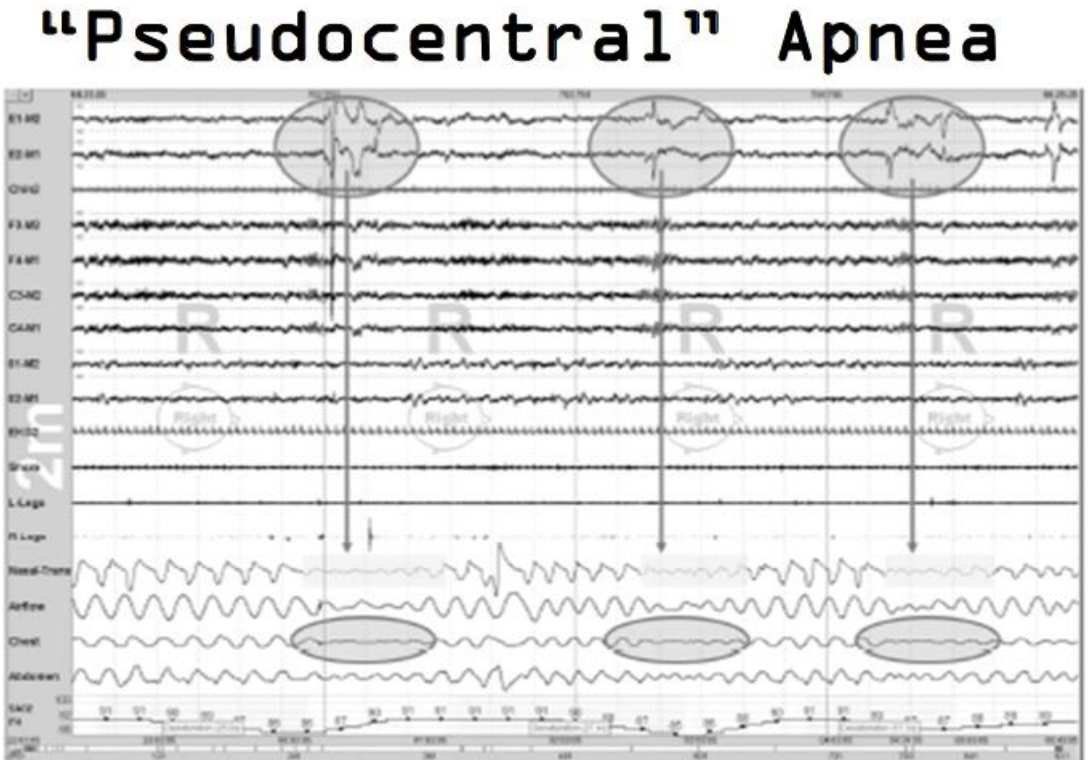
Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum



Source: Michael A. Grippi, Jack A. Elias, Jay A. Fishman, Robert M. Kotloff, Allan I. Pack, Robert M. Senior, Mark D. Siegel: *Fishman's Pulmonary Diseases and Disorders*:
www.accessmedicine.com

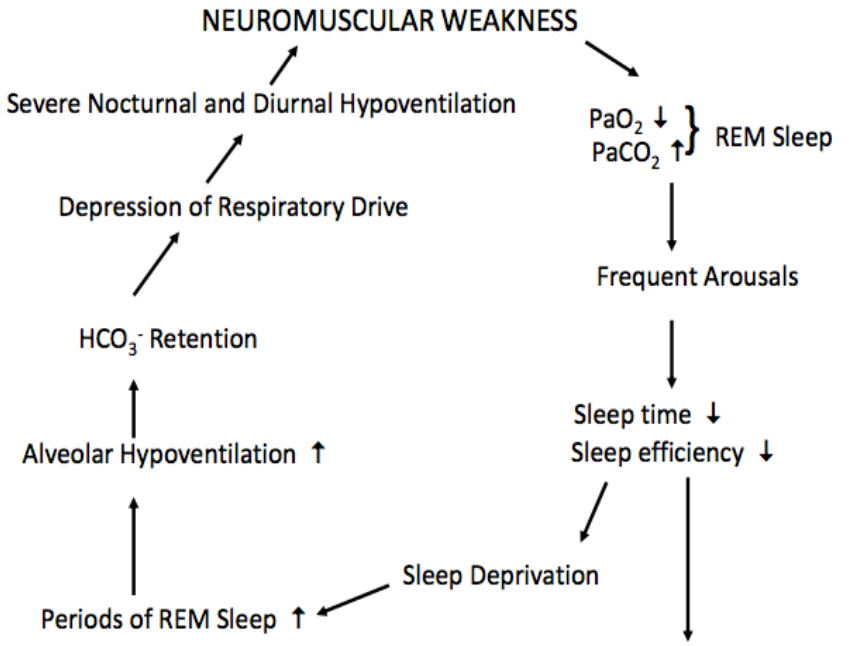
Sleep and NMD

- Suppression of intercostal muscle activity
- Shift of load to weak diaphragm
- More during phasic REM sleep
- More reduction in chest wall relative to abdominal muscles



(1)Aboussouan, AJRCCM Feb, 2015 (2) Nicolle MW. Semin Neurol 2009

Sleep and NMD



Summary of Changes

Perrin C et al. Muscle Nerve 29:5, 2004

Objectives

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

- Review of respiratory neuroanatomy
- General respiratory issues in neuromuscular disease (NMD)
- Sleep and neuromuscular disease
- **Specific Diseases**
- Respiratory evaluation of patients with NMD
- Management

Specific NMD

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

- Duchene Muscular Dystrophy (DMD)
- Spinal Muscle Atrophy
- Myotonic Dystrophy
- ALS

Duchene Muscular Dystrophy (DMD)

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

- X-linked disorder
- Most common lethal muscular dystrophy
- Average ago of diagnosis 5 years old
- Wheelchair dependent by adolescence
- Dystrophin; a protein complex that connects the cytoskeleton of a muscle fiber to the surrounding extracellular matrix through the cell membrane.
- Mutations in the DMD gene alter reading frame and prevent expression of dystrophin protein => muscle fibers degenerate
- Skeletal, cardiac and respiratory muscles primarily affected

Duchene Muscular Dystrophy (DMD)

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

Natural disease history

- Loss of ambulation 9-11 yo
- Progressive muscle loss & weakness with eventual loss of arm use
- Restrictive lung disease =>Nocturnal hypercapnia with subsequent diurnal hypercapnia
- Cardiomyopathy
- Death late teens/early adulthood

Spinal Muscle Atrophy (SMA)

Medical Service Company

20th ANNUAL JEAN S. MARX Memorial Education Forum

Spinal Muscle Atrophy (SMA)

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

- A neurodegenerative disorder- Autosomal recessive
- Most common genetic cause of death in infants
- 4-10 : 100,000 live birth
- Motor neuron disease (degeneration of anterior horn cells in spinal cord and motor nuclei in lower brain stem)
- Progressive muscle weakness and atrophy
- Cognition in unaffected
- Various age of presentation

Spinal Muscle Atrophy (SMA)

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

- Paucity of spontaneous movement
- Severe proximal muscle weakness and hypotonia
- Frog leg position
- Various degrees of bulbar weakness-weak cry/oropharyngeal control
- Extraocular muscles sparred
- Striking discrepancy between high level of social interaction and lack of motor skills
- Natural history results in progressive muscle loss and wasting, profound respiratory weakness

Myotonic Dystrophy (MD)

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

- Autosomal Dominant disorder
- Skeletal muscle weakness
- Respiratory muscle weakness (less likely in type 2)
- Sleep disturbance

  (low orexin level => Daytime hypersomnolence)
- Muscle pain
- Cardiac and endocrine abnormalities
- Cognitive impairment

ALS

- Disease of upper and lower motor neurons
- Most cases are sporadic
- < 5% familial

Clinical symptoms

- Vary Widely
- Respiratory and bulbar muscles are frequently involved
- Hypoventilation is predominant especially during sleep
- NPPV has been showed to improve survival and QOL

Aboussouan et al. Muscle Nerve 2001; 24

Objectives

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

- Review of respiratory neuroanatomy
- General respiratory issues in neuromuscular disease (NMD)
- Sleep and neuromuscular disease
- Specific Diseases
- **Respiratory evaluation of patients with NMD**
- Management

Respiratory Evaluation of Patients with NMD

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

Common Statements & Questions

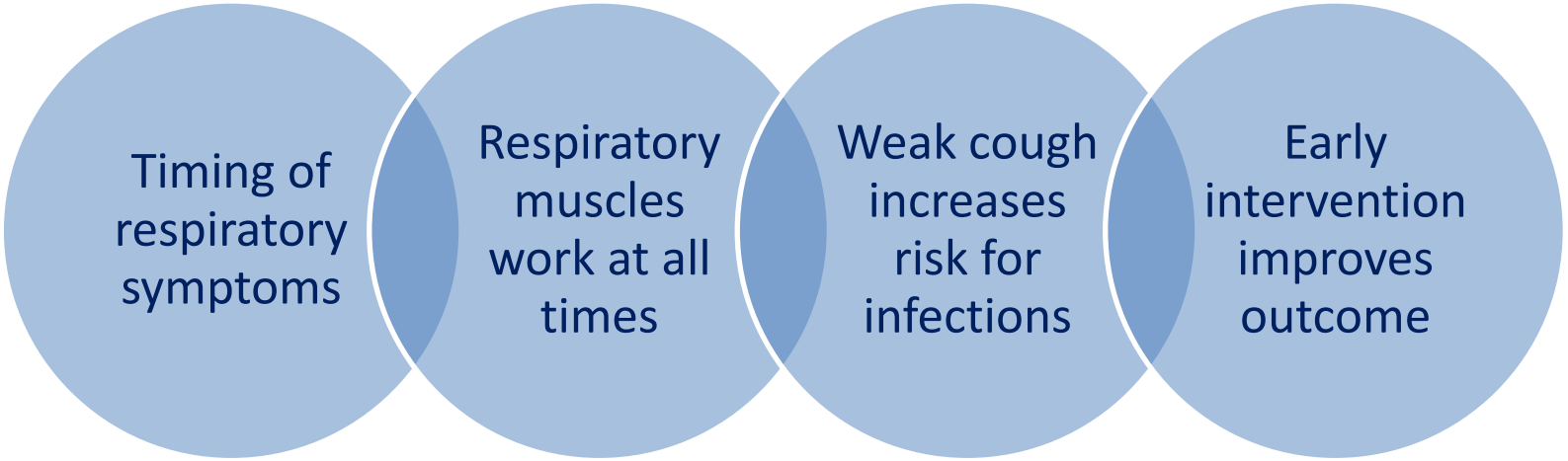
- I have no breathing problem
- I have no problems sleeping
- I don't cough frequently
- I didn't go to hospital for pneumonia or lung problems

Why do I need to see a lung doctor ???

Respiratory Evaluation of Patients with NMD

Medical Service Company

20th ANNUAL JEAN S. MARX Memorial Education Forum



Respiratory Evaluation of Patients with NMD

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

- Pulmonary Function Test
- Respiratory Muscle Forces
- Cough Effectiveness
- Sleep Studies
- Arterial Blood Gases
- Chest Radiography

Respiratory Evaluation of Patients with NMD

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

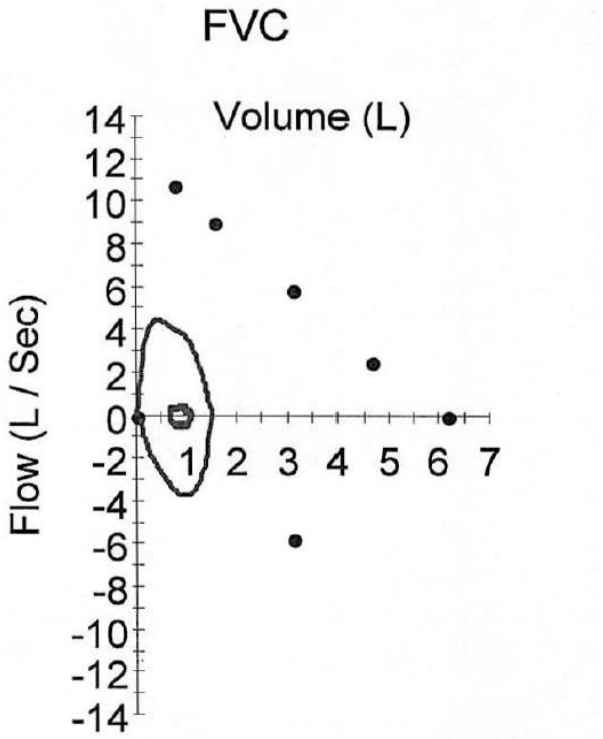
Pulmonary function tests

- No single abnormality is diagnostic
- Diagnosis is based on constellation of abnormalities
- Consider measuring FVC and VC in supine position (>10% reduction than upright position)
- Findings:
  - Restrictive pattern
  - Reduced maximum voluntary ventilation (MVV)
  - Reduced maximum inspiratory pressure (MIP) and maximum expiratory pressure (MEP)
  - Normal diffusion in absence of pulmonary parenchymal or vascular disease

Respiratory Evaluation of Patients with NMD

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

	Pre-Bronch		
	Pred	Actual	%Pred
SPIROMETRY			
FVC (L)	6.18	1.50	24
FEV1 (L)	5.12	1.50	29
FEV1/FVC (%)	84	100	119
FEV3 (L)		1.50	
FEV3/FVC (%)		100	
FEF 25-75% (L/sec)	5.27	3.86	73
FEF 75-85% (L/sec)		2.51	
FEF 50% (L/sec)	5.89	4.02	68
FEF 75% (L/sec)	2.47	3.10	125
FEF 200-1200 (L/sec)		3.79	
FEF Max (L/sec)	10.77	4.45	41
Expiratory Time (sec)		6.52	
FIF 50% (L/sec)	5.78	3.58	61
MEP (cmH2O)	231	33	14
MIP (cmH2O)	-126	-47	37



Respiratory Evaluation of Patients with NMD

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

Assess for ineffective cough

- Peak cough flow (PCF)

  <160 L/min =>ineffective cough

  160-270 L/min => at risk for respiratory infection
- MEP < 60 cmH_2O suggests ineffective cough
- Expiratory cough flow tracing
- Clinical evaluation

(1) Sancho J, Am J Phys Med Rehabil 2004, (2) Bach JR, Chest 1997

Respiratory Evaluation of Patients with NMD

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

Arterial blood gases to evaluate for daytime hypercapnia

Polysomnography in selected patients to assess for sleep disordered breathing and nocturnal hypoventilation

(Don't delay the initiation of NIV if patient meets RAD criteria)

Home Sleep Apnea Test should not be used

Respiratory Evaluation of Patients with NMD

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

Assess the need for ventilatory support

- Marked ABG abnormalities
- Reduced FVC <50% or VC < 15-20 ml/kg or VC < 1 L
- MIP < -30 cmH_2O => high risk for hypercapnia
- MEP < 40 cmH_2O => ineffective cough
- The rule of 20/30/40 (VC/MIP/MEP)

Mehta S, Respir Care 2005

Lawn ND, Arch Neurol, 2001

Objectives

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

- Review of respiratory neuroanatomy
- General respiratory issues in neuromuscular disease (NMD)
- Sleep and neuromuscular disease
- Specific Diseases
- Respiratory evaluation of patients with NMD
- **Management**

Management of Patients with NMD

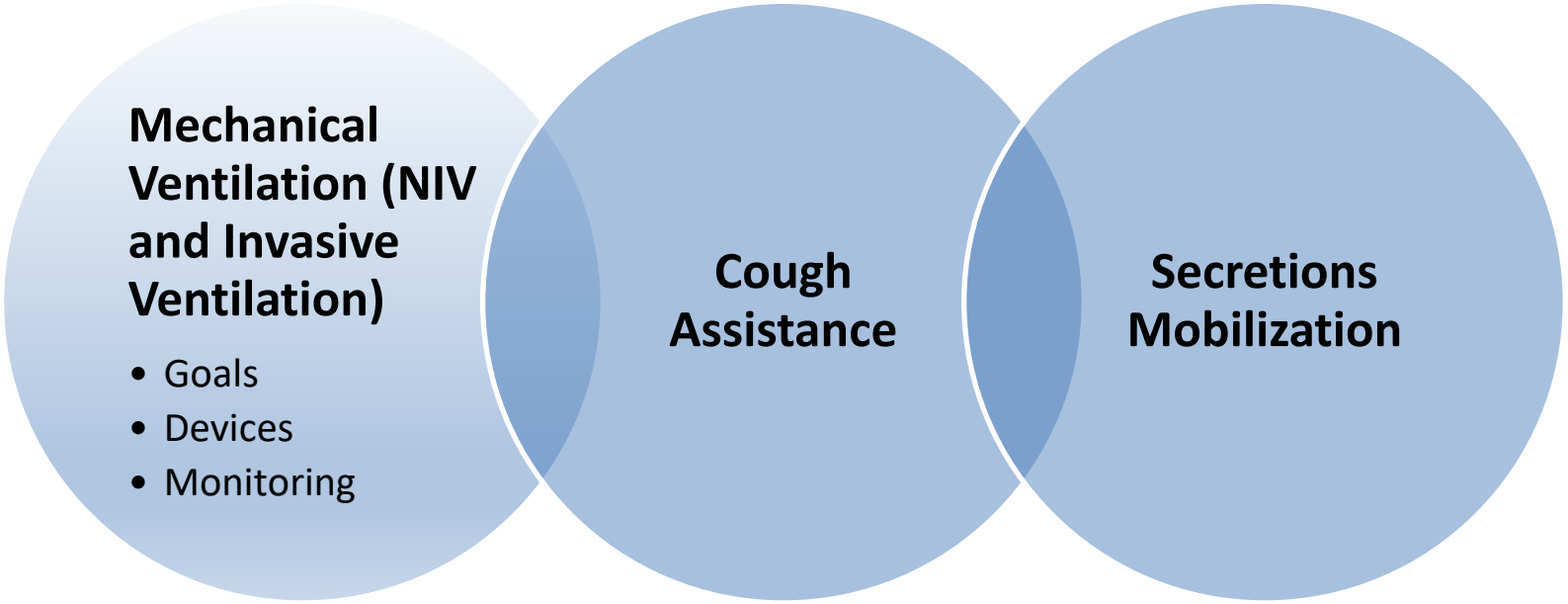
Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

Goals of Management

- Improve and stabilize gas exchange
- Ameliorate symptoms
- Improve sleep quality
- Improve quality of life
- Extend survival

Management of Patients with NMD

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum



Management of Patients with NMD

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

Non-Invasive Ventilation (NIV)

- Prevent or delay progression of chronic respiratory failure
- Prolong survival especially in patients with ALS
  - 92 patient with ALS
  - NIV vs. Standard care (after developing orthopnea, MIP<60%, or hypercapnia)
  - Improved quality of life and median survival of 205 days
  - NIV improves survival and quality of life in patients without severe bulbar dysfunction
  - NIV improves sleep-related symptoms but unlikely survival with severe bulbar symptoms

Bourke SC, Lancet Neurol. 2006

Management of Patients with NMD- NIV

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

Effects on Sleep:

	Before NPPV	With NPPV
RDI	10.5	3.1
REM RDI	20.5	3.0
Arousal Index	20.6	10.2
Light sleep	55%	44%
Slow-wave sleep	24%	34%
REM-sleep	18%	20%

Mellies et al ERJ 2003

Management of Patients with NMD-NIV

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

Important Considerations

- Bilevel support is the most common type- CPAP is not appropriate in most cases
- Back up rate is often recommended due to frequency of central and pseudocentral apnea
- Volume assured ventilation it theoretically better for progressive disease
- Proper mask interface is essential for successful treatment
- Don’t delay NIV for PSG
- In lab PAP titration may not be required in many cases
- Consider desensitization techniques to improve tolerance and adherence
- Home monitoring for therapy

Management of Patients with NMD- NIV

Medical Service Company

20th ANNUAL JEAN S. MARX Memorial Education Forum

Technical and Practical Aspects

Respiratory Assist Device

Mechanical Ventilator

Management of Patients with NMD- NIV

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

Respiratory Assist Devices	Mechanical Ventilators
Codes -E0470- Bilevel (S mode) -E0471- Bilevel (ST mode)	Codes -E0464
Features -Light weight -No mandatory alarm -Can be remotely monitored and controlled -Integrated humidity	Features -Heavy -Internal batteries -Can be remotely monitored -Multiple settings (day/night) -Alarms and monitors

Management of Patients with NMD- NIV

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

CMS Criteria for RAD

I. Restrictive Thoracic Disorders

Documentation of neuromuscular disease or severe thoracic cage abnormality in the patient's medical record →

Perform one of the following:

- **ABGs** (done while awake and on prescribed FiO_2) $PaCO_2 \geq 45$ mm Hg or
- **Sleep oximetry**
  Oxygen saturation ≤ 88% for ≥ 5 minutes, minimum 2 hours of recording time (on patient's prescribed FiO_2) or
- **For neuromuscular disease only:**
  Either FVC < 50% of predicted or MIP < 60 cm H_2O

→

COPD does not contribute significantly to pulmonary limitation →

(E0470) or (E0471)
Based on the treating physician's judgment

E0470 => BPAP- S (without back up Rate)
E0471 => BPAP- S/T or VPAPS

Management of Patients with NMD-NIV

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

Ventilators with Mask Interface:

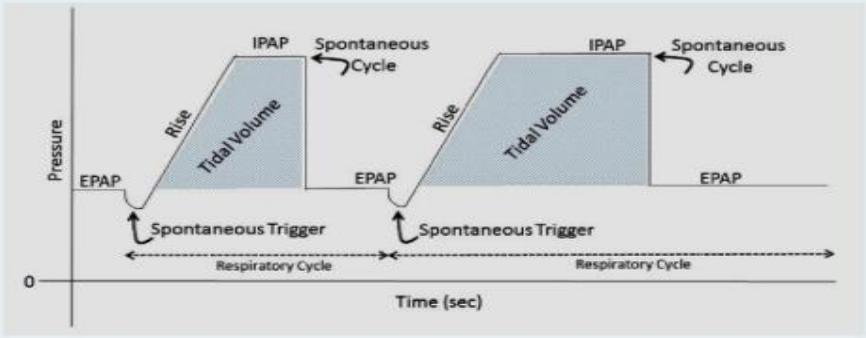
- Elevated PaCO2 despite the use of RAD
- Prolonged use of device (>8-12 hours daily)
- High pressure requirement needed for daytime support
- Portability is required

Management of Patients with NMD-NIV

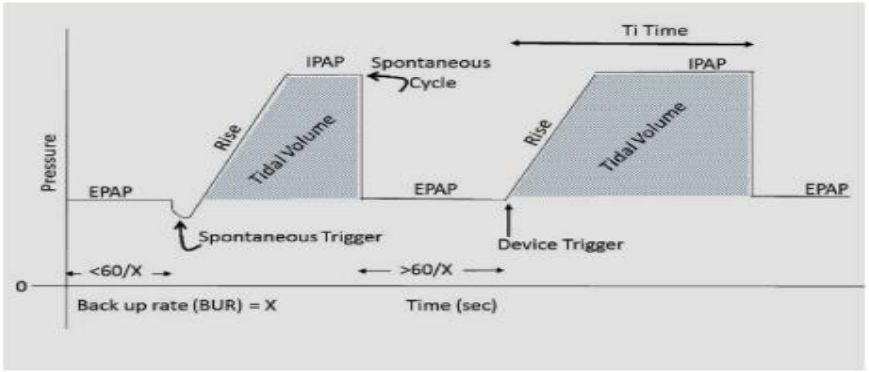
Medical Service Company

20th ANNUAL JEAN S. MARX Memorial Education Forum

S Mode (spontaneous)



ST mode (spontaneous/timed)
BIPAP-ST (Respironics)
VPAP-ST (ResMed)

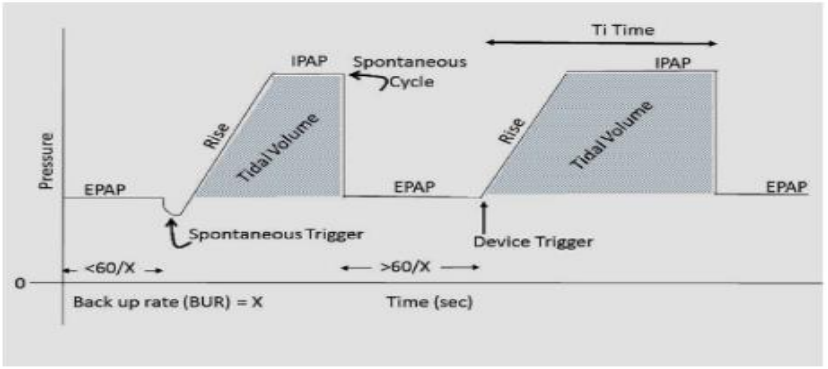


Management of Patients with NMD-NIV

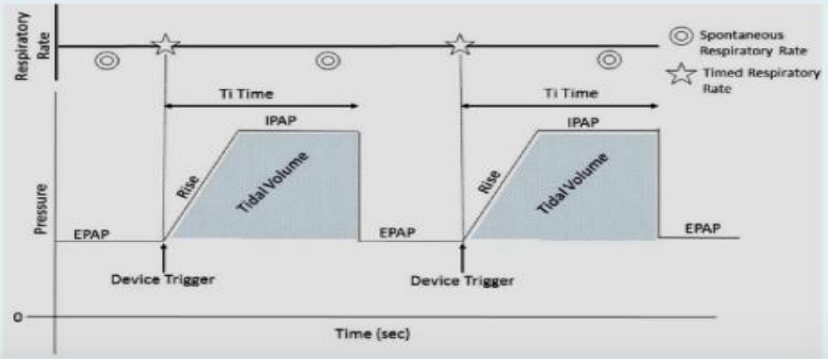
Medical Service Company

20th ANNUAL JEAN S. MARX Memorial Education Forum

ST mode (spontaneous/timed)
BIPAP-ST (Respironics)
VPAP-ST (ResMed)



T mode (timed)



Management of Patients with NMD-NIV

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

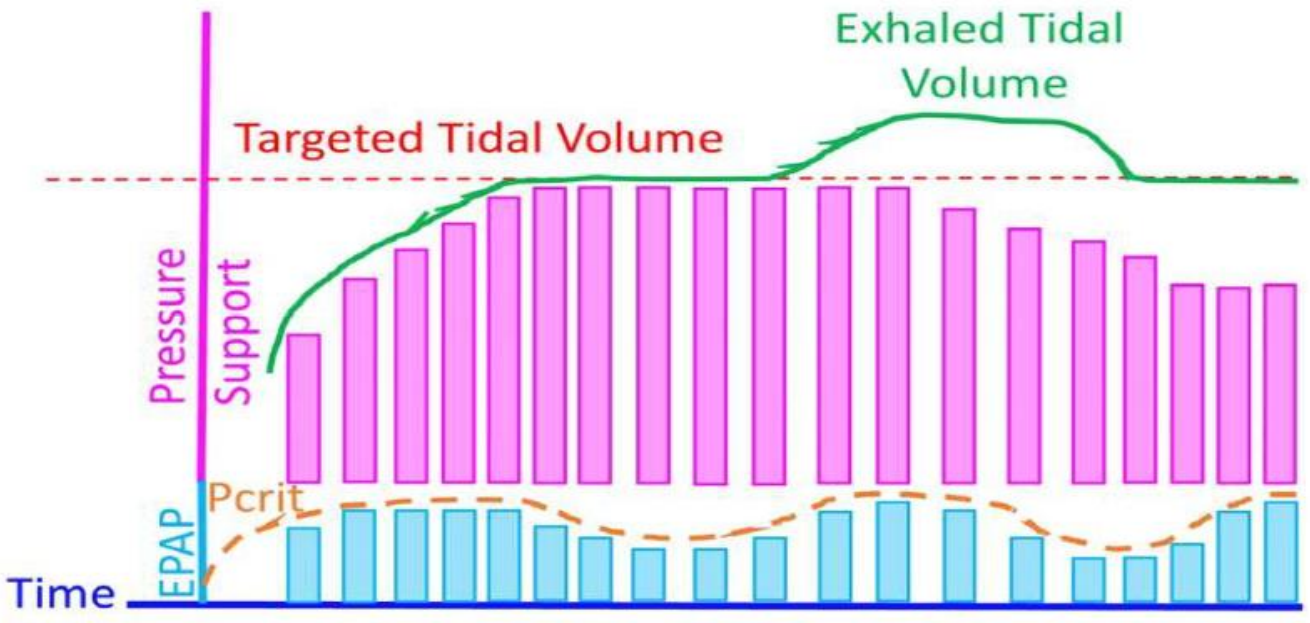
Volume Assured Pressure Support Modes

AVAPS (AE)	iVAPS (AE)
ST mode (Philips)	ST mode (ResMed)
Back up rate (Fixed or Auto)	Back up rate (intelligent)
Tidal Volume	Alveolar Ventilation (Va)
EPAP (fixed or auto)	EPAP (fixed or auto)
Remote monitoring	Remote monitoring

Management of Patients with NMD-NIV

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

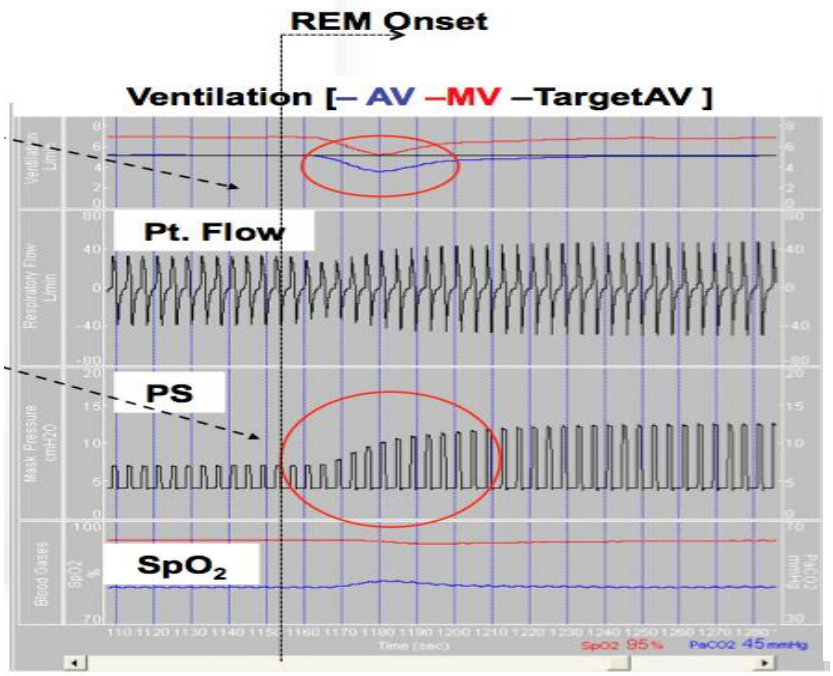
Volume Assured Modes with Floating EPAP (AVAPS-AE)

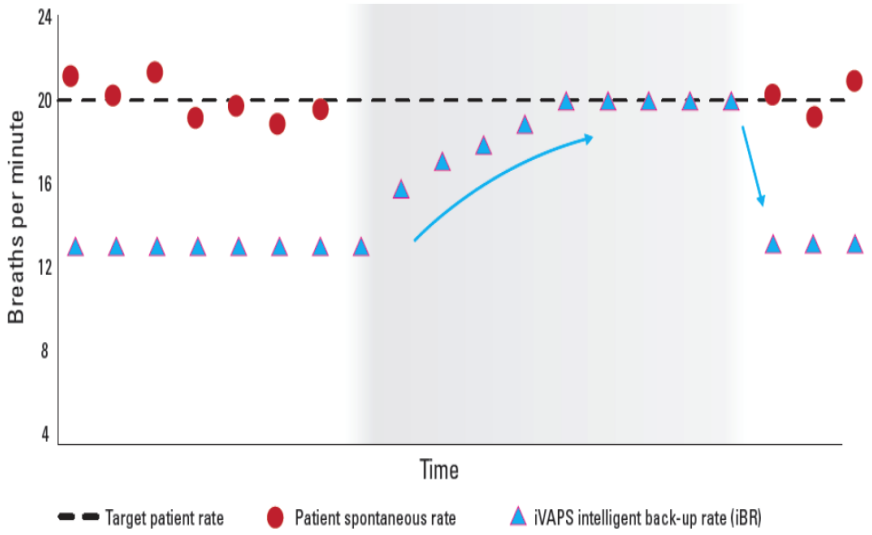


Management of Patients with NMD-NIV

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

Volume Assured with intelligent BUR (iVAPS)





Management of Patients with NMD-NIV

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

Volume Assured with intelligent BUR (iVAPS)

RR	10	12	15	20	24
Vt (mls)	690	595	500	405	358
MV (l/min)	6.9	7.14	7.5	8.1	8.58
Va (l/min)	5.7	5.7	5.7	5.7	5.7

Management of Patients with NMD-NIV

Medical Service Company

20th ANNUAL JEAN S. MARX Memorial Education Forum

Thorax August
2012 Vol 67 No 8

Editorial

Target volume settings for home mechanical ventilation: great progress or just a gadget?

Wolfram Windisch, Jan Hendrik Storre

cases patients were already familiar with pressure-preset NPPV prior to randomisation, thus indicating a selection bias. Therefore, the evidence for target volume setting remains inconclusive, and it is yet to be established whether these hybrid modes of ventilation have benefits that are clear and consistent enough to warrant official recommendations.

Murphy and colleagues randomly allocated 50 NPPV-naive patients with severe OHS (body mass index 50±7 kg/m^2)

Management of Patients with NMD-NIV

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

Why do we use VAPS?

- Most labs are not aggressive in initial titration
- Pressure requirements change with progression of disease
- Patient may not have access to in lab titration
- Advancing therapy without lab titration may speed the process and reduce paperwork
- VAPS improved compliance in some studies

Management of Patients with NMD-NIV

Medical Service Company

20th ANNUAL JEAN S. MARX Memorial Education Forum

Mask Interface – Full Face Masks

Management of Patients with NMD-NIV

Medical Service Company

20th ANNUAL JEAN S. MARX Memorial Education Forum

Mask Interface –Nasal Masks

- Reduces aerophagia
- Allows easy oral suctioning
- Easy to call for help
- Reduces claustrophobia

Management of Patients with NMD-NIV

Medical Service Company

20th ANNUAL JEAN S. MARX Memorial Education Forum

Mouthpiece Ventilation (SIP)

Management of Patients with NMD-NIV

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

Monitoring of NIV:

- Downloads from devices
- CO2 monitors
- Overnight Oximetry
- ABG
- Monitor for complications

Management of Patients with NMD-NIV

Medical Service Company

20th ANNUAL JEAN S. MARX Memorial Education Forum

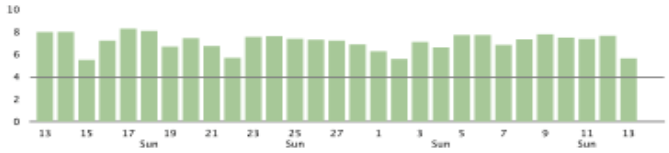
Compliance Report

Usage	02/13/2018 - 03/14/2018
Usage days	**29/30 days (97%)**
>= 4 hours	**29 days (97%)**
< 4 hours	**0 days (0%)**
Usage hours	206 hours 38 minutes
Average usage (total days)	6 hours 53 minutes
Average usage (days used)	7 hours 8 minutes
Median usage (days used)	7 hours 20 minutes

VPAP ST-A	
Serial number	22161395802
Mode	iVAPS
Target Alveolar Ventilation (Va)	4.5 L/min
EPAP	10 cmH2O
Min PS	4 cmH2O
Max PS	20 cmH2O
Target Patient Rate	18 bpm

Therapy						
Leaks - L/min	Median:	**0.0**	95th percentile:	**15.0**	Maximum:	**44.4**
Events per hour	AI:	**0.4**	HI:	**0.4**	AHI:	**0.8**

Usage - hours



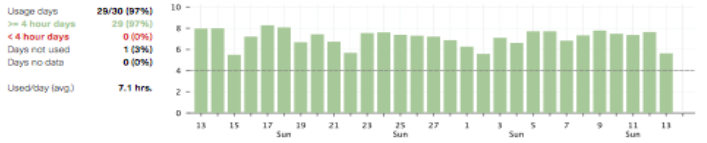
AirView™

Age: 28 years

Therapy Report

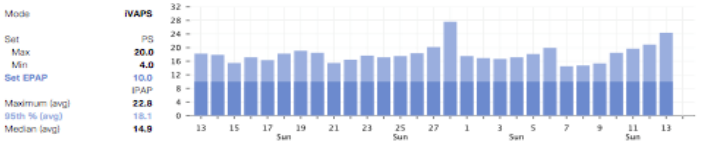
VPAP ST-A — SN: 22161395802

Usage (hours)

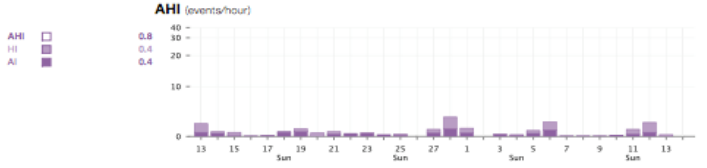


Leak (L/min)

Pressure (cmH2O)



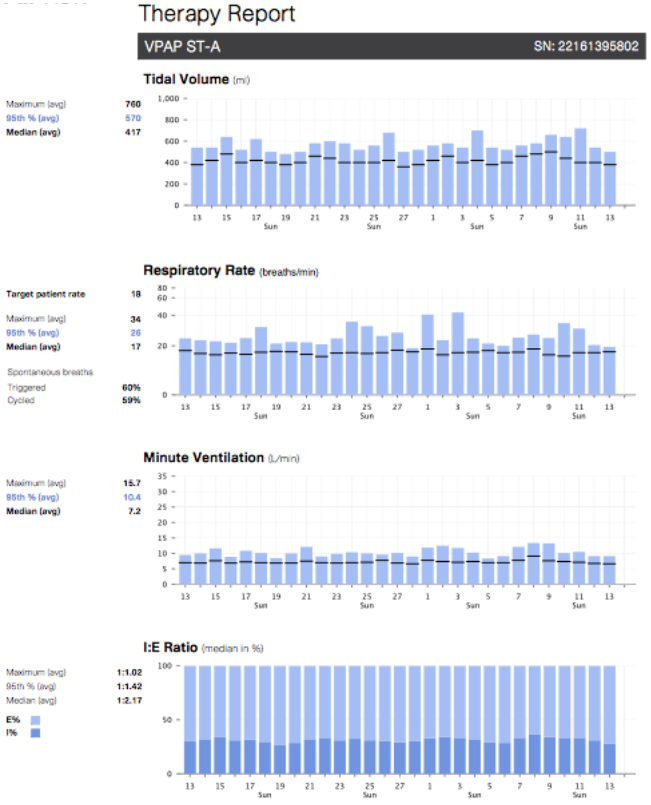
AHI (events/hour)



Management of Patients with NMD-NIV

Medical Service Company

20th ANNUAL JEAN S. MARX Memorial Education Forum



Management of Patients with NMD-NIV

Medical Service Company

20th ANNUAL JEAN S. MARX Memorial Education Forum

Compliance Report: Program 1

Usage: Program 1	03/20/2018 - 04/18/2018
Usage days	**30/30 days (100%)**
>= 4 hours	**30 days (100%)**
< 4 hours	**0 days (0%)**
Usage hours	311 hours 39 minutes
Average usage (total days)	10 hours 23 minutes
Average usage (days used)	10 hours 23 minutes
Median usage (days used)	10 hours 32 minutes

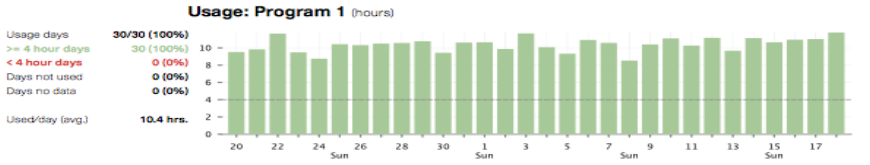
Astral 150: Program 1	SN: 22171124138
Device Settings: Program 1	
Mode	iVAPS
Circuit	Single with leak
Patient Interface	Mask
Mask	Pillows
Patient Type	Adult
Height	63.0 in
Inspiratory Phase Delivery Settings: Program 1	
Target alveolar ventilation	4 L/min
Min PS	4 cmH2O
Max PS	20 cmH2O
Rise time	450 ms
Inspiratory Trigger Settings: Program 1	
Target patient rate	15 breaths/min
Trigger	Medium
Inspiratory Phase Duration Settings: Program 1	
Ti Min	1.0 sec
Ti Max	2.2 sec
Cycle	25.0 %
Expiratory Phase Settings: Program 1	
EPAP	6 cmH2O

Therapy: Program 1						
Leak - L/min	5th %:	**1.1**	Median:	**4.8**	95th %:	**13.4**
Events per hour	AHI:	**0.4**	AI:	**0.0**	HI:	**0.4**

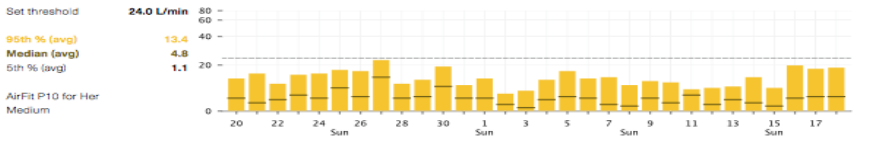
Therapy Report: Program 1

Astral 150 SN: 22171124138

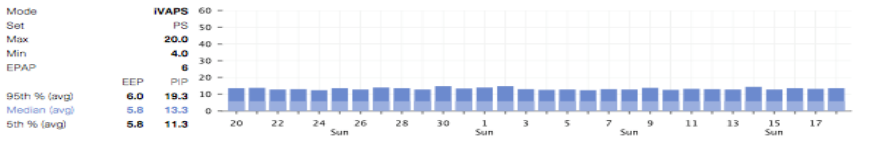
Usage: Program 1 (hours)



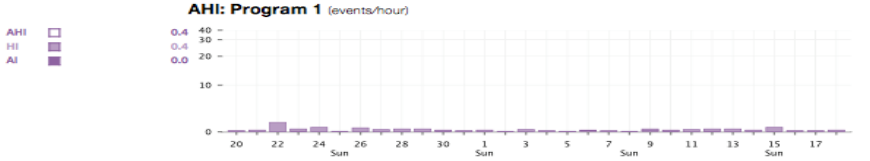
Leak: Program 1 (L/min)



Pressure: Program 1 (cmH2O)



AHI: Program 1 (events/hour)



Management of Patients with NMD-NIV

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

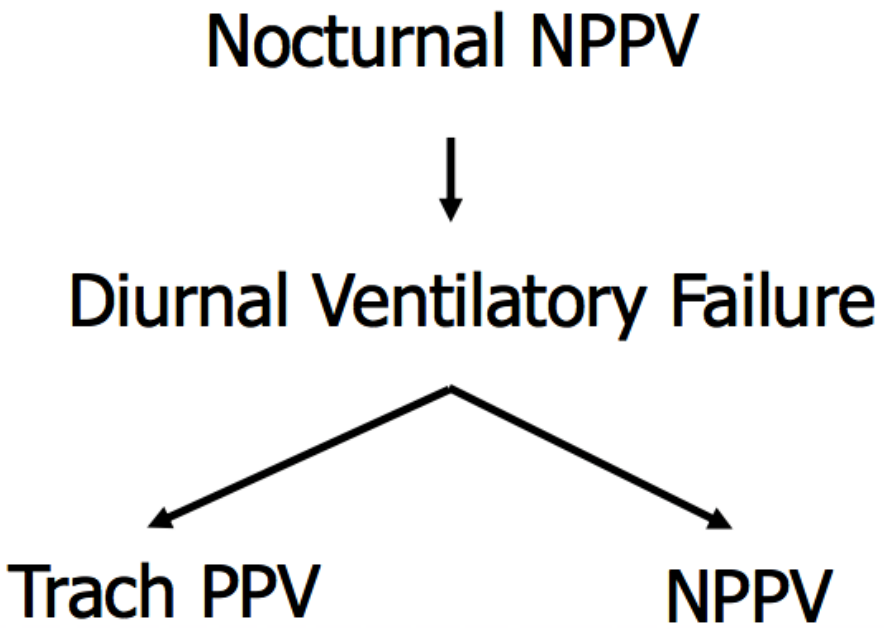
When to consider daytime NIV?

- Elevation of PaCO2 despite adequate treatment at night
- Daytime symptoms of dyspnea
- Frequent pneumonia due to cough insufficiency
  - Mouthpiece ventilator can improve cough

Management of Patients with NMD

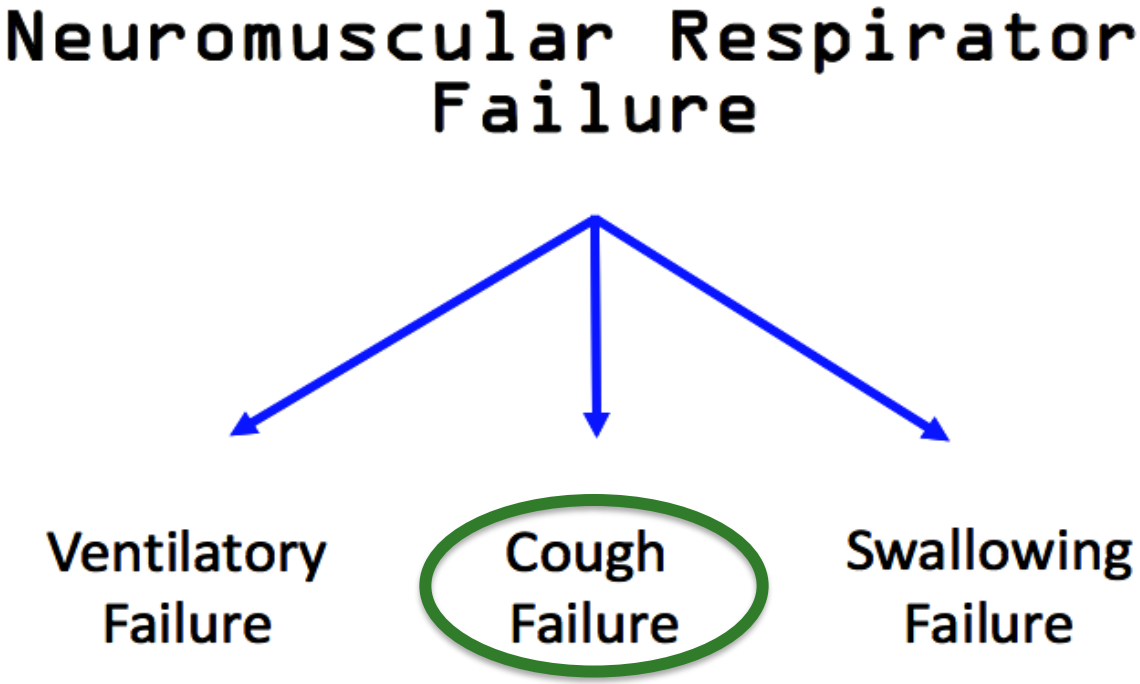
Tracheostomy

- Difficulty clearing secretions
- NIV is no longer sufficient
- Failure to wean off MV after an acute illness
- Discuss goals of care



Management of Patients with NMD

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

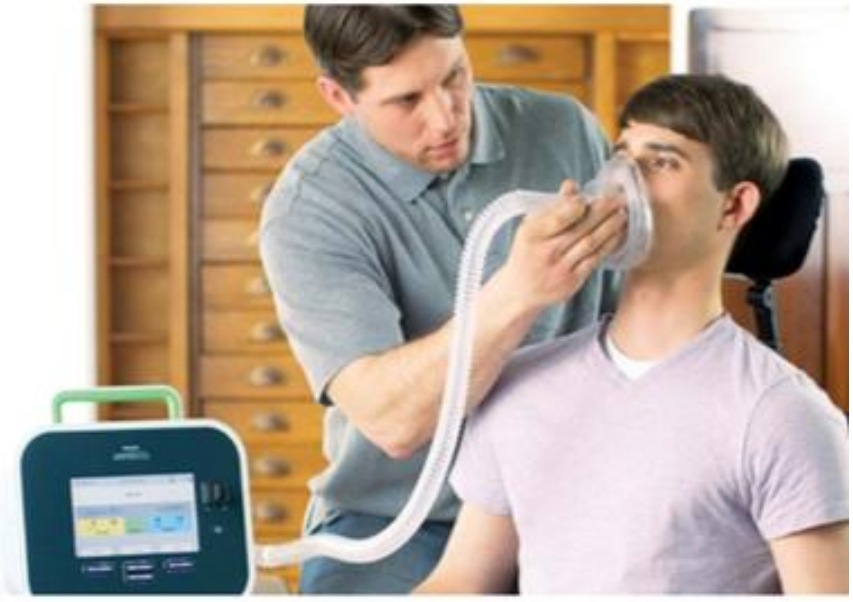


Management of Patients with NMD

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

Cough Assist Device

Mechanical insufflation-exsufflation



Management of Patients with NMD

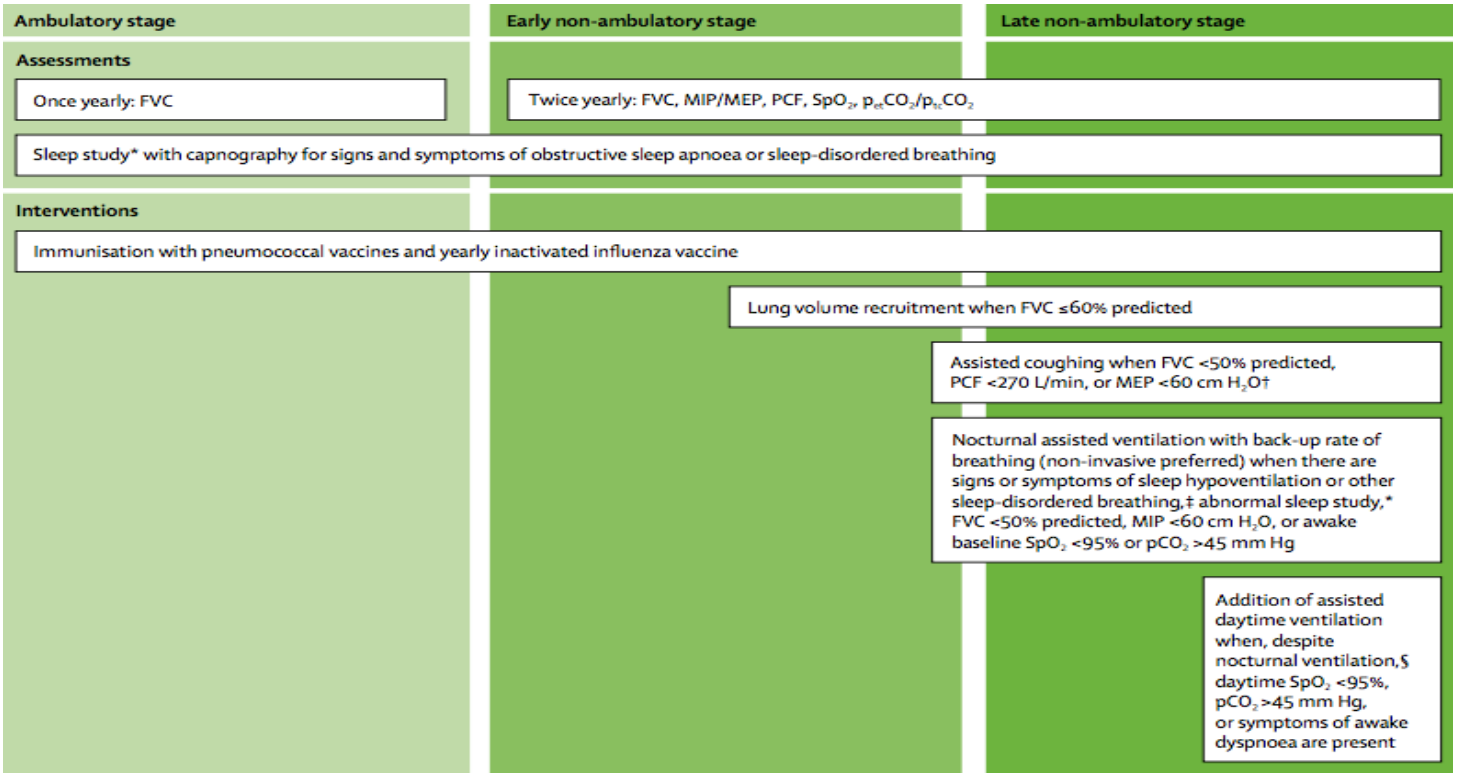
Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum

Secretions Mobilization Techniques

- High frequency chest wall oscillation
- Intrapulmonary percussive ventilation

Management of Patients with NMD DMD – Updated guidelines

Medical Service Company | 20th ANNUAL JEAN S. MARX Memorial Education Forum



David Birnkarnt, The Lancet Neuro April 2018

Medical Service Company

20th ANNUAL JEAN S. MARX Memorial Education Forum

Thank You