AFCH NEUROMUSCULAR DISORDERS (NMD) PROTOCOL

A. Definition of Therapy:

1. *Cough machine*: 4 sets of 5 breaths with a goal of I:E pressures approximately the same of 30-40. Inhale time = 1 second, exhale time = 1second, pause time = 2 seconds.
   If the patient is ≥ 6 years of age, attempt to use a mouthpiece, otherwise, use a mask.
   If the patient is less than 1 year of age, decrease pause time to 1 second.
2. *Airway Clearance Therapy (ACT)*
   a. *Chest Physical Therapy (CPT)*: side to side with HOB down.
   b. Intrapulmonary Percussive Ventilation (IPV): adjust frequency as tolerated using normal saline solution nebulized.
3. *Postural drainage*: supine with head of bed (HOB) down for 15 minutes or as tolerated.

B. Assessment

1. Patient assessment will be performed upon admission and daily 1 hour after the morning therapy using the triage scoring tool.
2. All assessments are to be done on room air and without respiratory support. If unable to score the patient off respiratory support, the patient immediate triages to total points of >16.
3. Triage scoring (total points) will determine the frequency. See Frequency Guidelines.
4. When the triage score is 0-4:
   a. If the patient is on home therapies, assess the patient every day.
   b. If the patient has no home therapies, assess the patient every shift for 48 hours.

C. Oxygenation

If oxygen saturation drops to < 93% acutely, perform cough machine and assess if the patient needs an extra treatment. The cough machine can be used as often as needed to clear lower airway secretions. If on room air (RA) and SpO2 remains ≤93%, apply respiratory support device (BiPAP or ventilator). If SpO2 remains ≤92%, apply supplemental oxygen to respiratory support device and reassess frequency of therapy. If patient is requiring supplemental oxygen, oxygen should be added to the cough machine.

D. Health link

Order will be written for "Respiratory Therapy per Protocol, protocol type Neuromuscular".

Name: Date:

NEUROMUSCULAR DISORDERS (NMD) TRIAGE SCORING

Assess on room air and without respiratory support upon admission and 1 hour after morning treatment. **If unable to assess off respiratory support, immediate triage to >16.**

Points	0	1	2	3	Total Points
Respiratory Rate 0-9years of age ≥10 years of age	 20-30 15-20	 31-40 21-30	 41-50 31-40	 >50 >40	
Heart Rate	80-100	100-120	120-140	>140	
RA Pulse Ox	>94%	94%-91%	90%-85%	<85%	
FI02	Room Air	≤lpm	1.1 lpm-3lpm	>3lpm	
Breath Sounds	Clear	Decreased Unilaterally	Crackles	Decreased Bilaterally	
Color	Normal	Pale	Cyanotic	Diffuse Cyanosis	
CXR (if none on day of assessment-continue with previous score)	Clear	Infiltrates atelectasis	Infiltrates more than 1 lobe or lobar collapse	Unilateral or bilateral whiteout	
WOB	Normal	Nasal Flaring	Retractions	Grunting and Flaring	
Secretions With cough machine.	None	Scant amounts	Moderate amounts	Copious amounts	
Mental State	Alert	Sleepy	Lethargic and arousable	Lethargic and not arousable	
Cough	None	Rare	Occasional	Frequent	

FREQUENCY GUIDELINES

Total Points	BiPAP	Treatments
≥16 points	Continuously	Every 2 hours (cough assist-ACT-cough assist-drainage-cough assist; oral suction as needed) Cough assist every 2 hours as needed
11-15 points	4 hours off twice daily	Every 4 hours (cough assist-ACT-cough assist-drainage-cough assist; oral suction as needed) Cough assist every 2 hours as needed.
5-10 points	at night only	4 times daily (cough assist-ACT-cough assist-drainage-cough assist; oral suction as needed)
0-4 points	Baseline	As needed (cough assist-ACT-cough assist-drainage-cough assist; oral suction as needed) or home therapy regimen

If treatment frequency is different than triage score indicate why.
____RCP discretion ____MD discretion Who and Why

I. Nonintubated Patient

A. Protocol

1. Initial Protocol
    a. Cough machine +/- manual cough assist per patient preference and tolerance, then orally suction.
    b. Airway clearance with IPV or CPT or vest
    c. Cough machine +/- manual cough assist per patient preference and tolerance, then orally suction.
    d. Postural drainage for 15minutes as tolerated.
    e. Cough machine +/- manual cough assist per patient preference and tolerance, then orally suction.
2. 48 Hours (2 days) of q2 hour therapy and no improvement based on assessment:
    a. Evaluate CXR with attending physician:
        i. If airspace disease present and/or airway clearance is not tolerated by patient, decrease frequency of therapy.
        ii. If atelectasis and mucus plugging present then therapy will be preformed every two hour. Change mode of airway clearance.
    b. Consider decreasing frequency during sleep with attending physician.
3. 72 Hours (3 days) of q2 hour therapy and no improvement based on assessment:
    a. Evaluate CXR with attending physician:
        i. If airspace disease present and/or airway clearance is not tolerated by patient, consider decreasing frequency of therapy.
        ii. If atelectasis and mucus plugging present then therapy will be preformed every two hour. Alternate modes of airway clearance.
    b. Consider decreasing frequency during sleep with attending physician.
4. 96 Hours (4 days) of q2 hour therapy and no improvement based on assessment:
    a. Evaluate CXR with attending physician:
        i. If airspace disease present and/or airway clearance is not tolerated by patient, decrease frequency of therapy to q3 hour.
        ii. If atelectasis and mucus plugging present alternate modes of airway clearance, decrease frequency of therapy to q3 hour.
    b. Consider decreasing frequency during sleep with attending physician.
5. 120 Hours (5 days) of q3 hour therapy and no improvement based on assessment:
    a. Evaluate CXR with attending physician:
        i. If airspace disease present and/or airway clearance is not tolerated by patient, decrease frequency of therapy to q4 hour.
        ii. If atelectasis and mucus plugging present alternate modes of airway clearance, decrease frequency of therapy to q4 hour.
        iii. Consider hypertonic saline 3% or Pulmozyme or Mucomyst.
        iv. Consider Bronchoscopy.
    b. Consider decreasing frequency during sleep with attending physician.

B. Post-operative Patient

Provide treatments within one hour after returning from surgery and continue every 4 hours until following am. Reassess in am for treatment schedule to return to baseline.

II. Intubated Patient

Intubated patients do not have assessments using the triage tool and fall into one of two categories; those intubated for post operative care and those intubated for respiratory failure.

A. Post-operative Patient

Provide treatments within one hour after returning from surgery and continue every 4 hours in the following sequence:

a. Cough machine set at inhale pressure +40 for 1 second, exhale pressure –40 for 1 second and 2 second pause, 4 sets of 5 breaths. Bleed in oxygen to cough machine if requiring supplemental oxygen.
b. Endotracheal tube and nasal suctioning.
c. Intrapulmonary percussive ventilation (IPV).
d. Cough machine set at inhale pressure +40 for 1 second, exhale pressure –40 for 1 second and 2 second pause, 4 sets of 5 breaths. Bleed in oxygen to cough machine if requiring supplemental oxygen.
e. Endotracheal tube and nasal suctioning.

B. Respiratory Failure Patient

1. Therapy will be performed every two hours in the following sequence:
   a. Cough machine set at inhale pressure +40 for 1 second, exhale pressure –40 for 1 second and 2 second pause, 4 sets of 5 breaths. Bleed in oxygen to cough machine if requiring supplemental oxygen.
   b. Endotracheal tube and nasal suctioning.
   c. Intrapulmonary percussive ventilation (IPV).
   d. Cough machine set at inhale pressure +40 for 1 second, exhale pressure –40 for 1 second and 2 second pause, 4 sets of 5 breaths. Bleed in oxygen to cough machine if requiring supplemental oxygen.
   e. Endotracheal tube and nasal suctioning.

2. 48 Hours post intubation and no improvement in ventilator settings or CXR:
   a. Evaluate CXR with attending physician:
      i. If airspace disease present and/or airway clearance is not tolerated by patient, consider decreasing frequency of therapy.
      ii. If atelectasis and mucus plugging present then therapy will be preformed every two hours in the following sequence:
         a. Cough machine set at inhale pressure +40 for 1 second, exhale pressure –40 for 1 second and 2 second pause, 4 sets of 5 breaths.
         b. Endotracheal tube and nasal suctioning.
         c. CPT or vest therapy (no vest for children under the of 2)
         d. Cough machine set at inhale pressure +40 for 1 second, exhale pressure –40 for 1 second and 2 second pause, 4 sets of 5 breaths.
         e. Endotracheal tube and nasal suctioning.
         f. Evaluate for drainage.
   b. Consider decreasing frequency during sleep with attending physician.

3. 96 Hours post intubation and no improvement in ventilator settings or CXR, therapy will be performed every two hours in the following sequence:
    a. Cough machine set at inhale pressure +40 for 1 second, exhale pressure –40 for 1 second and 2 second pause, 4 sets of 5 breaths.
    b. Endotracheal tube and nasal suctioning.
    c. Alternate IPV and CPT or vest therapy every other treatment.
    d. Cough machine set at inhale pressure +40 for 1 second, exhale pressure –40 for 1 second and 2 second pause, 4 sets of 5 breaths.
    e. Endotracheal tube and nasal suctioning.
    f. Evaluate for drainage.
    g. Recruitment Maneuver.
    h. Consider hypertonic saline 3% or Pulmozyme or Mucomyst.
    i. Consider Bronchoscopy.
    j. Consider decreasing frequency during sleep with attending physician.

C. Wean

1. Wean per the ventilator management protocols.

2. Work toward the following prior to extubation:
    a. Afebrile.
    b. Not requiring supplemental O2.
    c. CXR is without atelectasis or infiltrates.
    d. Off respiratory depressants.
    e. Airway suctioning is 3-4 times/8 hours or less.

D. Extubation

1. Extubate to continuous nasal ventilation and no supplemental O_2 such as BiPAP with settings IPAP 14-20 (15) and EPAP 3-6 (5) using the spontaneous timed (S/T) mode with a backup rate to match the patient's respiratory rate.

Age	**Respiratory Rate**	**Normal Predicted**	**Inspiratory Time (seconds)**
<2	30	20-25	.5-.8
2-5	20	15-20	.8-1
>5	16	12-15	1

Weaker patients > 2 years may require a higher respiratory rate.

2. If oxygen saturation drops to < 93% acutely, perform cough machine and assess if the patient needs an extra treatment. The cough machine can be used as often as needed to clear lower airway secretions. If on room air (RA) and SpO2 remains ≤93%, apply respiratory support device (BiPAP or ventilator). If SpO2 remains ≤92%, apply supplemental oxygen to respiratory support device and reassess

frequency of therapy. If patient is requiring supplemental oxygen, oxygen should be added to the cough machine.

3. Follow the normal therapy regime according to triage scoring and frequency guidelines. The goal is to use BiPAP per nasal mask while sleeping only.

E. Initial settings for specific device:

1. Trilogy ventilator passive circuit

a. ST or PC modes: Rate-age based, IPAP based on peak inspiratory pressure from mechanical ventilator and may require higher pressure depending on leak, PEEP 4, inspiratory time based on age, rise time 1. For optimal sensitivity set to flow trigger with flow trigger sensitivity 1 (most sensitive) to 4. If ST mode, set flow cycle sensitivity to 50% (most sensitive 90% - least sensitive 20%). The goal is to see chest wall rise with inspiration during sleep with minimal to no abdominal breathing.

b. Use nasal mask with exhalation ports.

d. Set disconnect alarm.

e. **Assess triggering and synchronization** and fine-tune sensitivity settings.

f. If on RA and SpO2 ≤93%: Increase the peak inspiratory pressure by 2 to a maximum of 20 cm H20.

2. Respironics V60 BiPAP

a. ST mode, IPAP 14-20, EPAP 4-6 with backup rate set to patient's normal predicted and inspiratory time set based on age. The goal is to see chest wall rise with inspiration during sleep with minimal to no abdominal breathing.

b. **Assess triggering and synchronization** and fine-tune adjustable settings.

c. If on RA and SpO2 ≤93%: Increase the peak inspiratory pressure by 2 to a maximum of 20cm H20.

III. **Post-operative Patient** – new tracheotomy

A. Provide treatments within one hour after returning from surgery and continue every 4 hours in the following sequence:

1. Cough machine set at inhale pressure +30 for 1 second, exhale pressure –30 for 1 second and 2 second pause, 4 sets of 5 breaths. Bleed in oxygen to cough machine if requiring supplemental oxygen.
2. Tracheostomy tube and nasal suctioning.
3. Manual chest physiotherapy every 4 hours.
4. Cough machine set at inhale pressure +30 for 1 second, exhale pressure –30 for 1 second and 2 second pause, 4 sets of 5 breaths. Bleed in oxygen to cough machine if requiring supplemental oxygen.
5. Tracheostomy tube and nasal suctioning

B. Decrease frequency of treatments as secretion volume decreases.

C. After first tracheostomy tube change, may add postural drainage following secretion mobilization technique with treatments.

Appendix: Abbreviation Table

+, positive
-, negative
ACT, airway clearance technique
BIPAP, Biphasic positive airway pressure
CXR, chest xray
EPAP, expiratory positive airway pressure
I:E, inspiratory to expiratory
IPAP, inspiratory positive airway pressure
LTV, lap top ventilator
ml/kg, milliliters per kilogram
NPPV, noninvasive positive pressure ventilation
O2, oxygen
RA, Room Air
SIMV/PC, synchronized intermittent mechanical ventilation with pressure control
SpO2, saturated partial oxygen concentration
S/T, spontaneous timed
VE, minute ventilation
WOB, work of breathing