CARE GUIDELINES FOR SMA CHILDREN WITH ACUTE RESPIRATORY ILLNESS

For the not yet intubated patient, perform the following a minimum of every 2-4 hours, or as frequently as needed if there are copious secretions. Alternate position of patient every cycle to either side or back, guided by x-ray appearance:

1. In-exsufflator cough machine set at inspiratory pressure of + 30 to 40 cm H20 for 1-2 seconds, expiratory pressure –30 to 40 cm H20 for 1 second and 1 to 2 second pause: 4 sets of 5 breaths, followed by

2. Upper airway suctioning (avoid deep suctioning when possible, as it can result in edema and worsen the situation)

3. If there is partial or complete collapse of a lobe, intubation may be inevitable, but could potentially be reversed with continuous BiPAP support (pressures 16-20 over 4-8) and frequent respiratory PT/cough assist treatments to remove upper airway secretions.

4. Oxygen supplementation can be used with BiPAP support, but oxygen without BiPAP support should be avoided, as it will result in decreased respiratory effort, atelectasis and CO2 retention. If work of breathing is too great, with HR persistently 10-20 % above baseline, intubation may be inevitable and is best done proactively.

5. When ill, patients are remarkably vulnerable to decompensation with respiratory treatments. Pre-treatment hyperventilation with bag and mask, and supplemental oxygen via cough assist tubing during treatments may be helpful. Be prepared to intubate as necessary; these patients have an increased propensity to demonstrate a vagal response with stress; this may resolve fairly quickly if the stimulation evoking it is stopped, and oxygen with or without bag/mask ventilation is given immediately.

6. If intubation is necessary, it is important to remember that these patients often have jaw contractures, which can make intubation a challenge.

Nutritional Management Guidelines:

It is common practice to stop feeds when monitoring these patients during illness due to the increased risk of aspiration should they require urgent intubation. However, these patients have little to no reserve of lean body mass to mobilize energy, fatigue more quickly when catabolic, and are more likely to become acidotic when adequate nutrition is lacking.

1. If fasting is expected to exceed 6 hours, recommend institution of continuous NJ feeds using an elemental or easy to digest formula containing protein.

2. If intubation seems imminent, and aspiration risk is considered too high, institute peripheral or total parenteral nutrition (PPN or TPN) with dextrose 10-15% and 1.5 grams/kg/24 hours amino acid with vitamins/electrolytes. Intralipid infusion can be used, but calories from lipid shouldn't exceed 15-20% of total calories.

3. Avoid giving just dextrose and electrolytes without amino acids for prolonged periods of time, as it frequently results in hyperglycemia and acidosis.

Revised 1/2007 by Kathryn J. Swoboda, M.D. Respiratory guidelines by Mary Schroth, M.D. Pediatric Pulmonology, University of Wisconsin. Adapted from Bach JR et al, Spinal muscular atrophy type 1: a noninvasive management approach. Chest 2000;117:1100.

For the intubated patient, performing the following a minimum of every 4 hours (alternate position of patient every cycle to either side or back, guided by x-ray appearance):

7. Intrapulmonary percussive ventilation followed by
8. In-exsufflator cough machine set at inspiratory pressure of + 30 cm H20 for 1 second, expiratory pressure –30 cm H20 for 1 second and 1 to 2 second pause: 4 sets of 5 breaths, followed by
9. Endotracheal tube and airway suctioning

Note: the inexsufflator machine can be used as often as needed followed by endotracheal suctioning to help remove excessive lower airway secretions

Extubate when patient is:

1. Afebrile
2. **not requiring additional supplemental O2**
3. **CXR without atelectasis or infiltrates**
4. **off respiratory depressants for at least 24 hours – yes, this includes ativan, versed, valium and derivatives**
5. necessity of airway suctioning is close to baseline

Extubate to continuous nasal or mask ventilation (without supplemental 02 or minimal supplemental O2) such as BiPAP, settings: IPAP 12 to 20, EPAP: 3 to 6 using the spontaneous mode. Use ST (spontaneous timed) mode with backup rate if patient is unable to initiate a breath with BiPAP. Backup rates are often higher than in patients without neuromuscular disease. Use oximetry as a guide for use of expiratory aids, postural drainage and CPT.

Following extubation perform the following at least every 4 hours:

1. Chest PT manual or IPV, followed by
2. In-exsufflator cough machine, set at inspiratory pressure of +40 for 1 second, expiratory pressure –40 for 1 second and pause for 1 to 2 seconds, 4 sets of 5 breaths, followed by
3. Postural drainage (trendelenberg) up to 15 minutes as tolerated, followed by
4. In-exsufflator cough machine, 4 sets of 5 breaths, settings as in 2.

The inexsufflator cough machine can be used as often as every 10 minutes to clear lower airway secretions. Use acutely if oxygen saturation drops to < 94%. Wean from BiPAP during the day as tolerated. Goal is to use BiPAP per nasal mask while sleeping only, or to wean to their prior baseline use of BiPAP. Wean airway clearance regimen above as airway secretions decrease working toward a QID schedule or less.

Nutritional Management during concomitant illness/catabolic states is critical! Do not allow fasting > 6 hours in SMA type 1 subjects, or > 8 to 12 hours in SMA type 2 subjects or other neuromuscular patients. SMA patients have a secondary fatty acid oxidation defect and virtually all have reflux, delayed gastric emptying and autonomic gastrointestinal dysfunction which worsens with illness: recommend immediate institution of continuous feeds via NJ, plus/minus supplementation with PPN including Dextrose 10-15%, approximately 1.5 gms/kg amino acids per 24 hour period and no more than 10-15% intralipid if indicated.

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