

Emergency Care

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Parent JOIN THE FIGHT.
Project END DUCHENNE.
Muscular
Dystrophy

Emergency Care



- Emergencies never happen when or where you would like them to happen
 - Most likely NOT when families are not close to a center/medical provider familiar with Duchenne
- Parents may be the experts in the room
- **What do parents need to know?**

Helpful to Have



- Medical alert jewelry (for first responders)
 - Name, DOB, phone, NKDA (or allergies)
 - Medications
 - NO INHALED ANESTHESIA
- Copy of your last note(s)
- Phone numbers of your NMS/subspecialists
- All equipment/medications
 - Pressures used

PPMD Website



- Emergency Care Information and Resources
 - Emergency Card
 - Mobile app
 - Safe/unsafe anesthesia
 - O2 recommendations/precautions
 - FES
 - Fractures

Respiratory Emergencies

- URI and flu especially bad this year
 - Ability to breathe
 - O₂/CO₂ levels
- Blog: *Avoiding the Emergency Room*
 - Links to several resources
- Bring with you
 - Equipment
 - Medications
 - Insist they be used!



Managing Colds with Duchenne

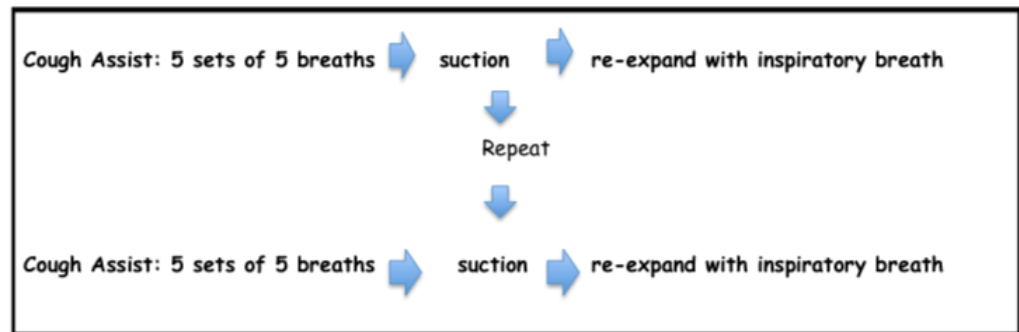
Keeping Your Lungs Healthy When You Have Breathing Muscle Weakness

Handling Colds

During a cold, a weak cough can become weaker, making it more difficult to clear mucus from your lungs. People with a muscular dystrophy have normal lungs. **If your oxygen saturation is going down, it means you need to cough to clear the mucous so that you can breathe more easily.** Giving oxygen will not help raise your oxygen saturation.

An oxygen saturation of 96% is normal; an oxygen saturation level of 95% indicates that your lungs are retaining carbon dioxide; an oxygen saturation level of 94% indicates further carbon dioxide retention. The following steps are very helpful in keeping your lungs clear and keeping oxygen saturations normal (above 96%).

1. At least every 4 hours:



2. Use the Cough Assist whenever you hear or feel a rattle in the chest. Suction the mouth when mucus is stuck in the back of throat.
3. If you have them at home, use BIPAP or the ventilator with all sleep (i.e., naps and overnight). With colds, we all are weaker.
4. Use the oximeter, once a day when well and at least 3 times a day when sick. **If the oxygen saturation is less than 96%, use the Cough Assist to clear secretions and then recheck the oxygen saturation. If the oxygen saturation remains at 96% or less, the Cough Assist is not helping to increase the oxygen saturation, Please call your primary care provider, pulmonologist or seek medical attention.**

Assisted Cough

One Page History

About Me

Hi! My name is:

I have:

When I have a respiratory infection or pneumonia, my weak cough makes it difficult to keep mucus from building up in my lungs. I use a Cough Assist Device to help bring up the mucus.

The settings for my Cough Assist are:

Inspiratory Pressure: cm H₂O

Expiratory Pressure: cm H₂O

Inspiratory time: sec; Expiratory time: sec; Pause: sec;

If my oxygen saturations are less than 94% then I need more frequent Cough Assist and possibly BIPAP (IPAP 12-20 cm H₂O; EPAP 3-6 cm H₂O) or nasal mask non-invasive ventilation (assist controlled volume ventilation).

*Using just oxygen to treat low oxygen saturations can mask and cause CO₂ retention and respiratory acidosis! **As a general rule, oxygen without BIPAP should be avoided!***

BIPAP and nasal mask non-invasive ventilation can reverse hypoxemia and respiratory acidosis AND prevent the need for intubation.

My Muscular Dystrophy Neurologist is:

Contact #:

Pulmonologist is:

Contact #:

Cardiologist is:

Contact #:

Respiratory Therapist is:

Contact #:

Oxygen



- Two parts to breathing
 - Mechanics of breathing in and out
 - Oxygen/carbon dioxide exchange
- Breathing with Duchenne
 - Diaphragm/intercostal muscles are weak
- Delicate oxygen/CO₂ balance
 - Have enough oxygen
 - No need to breath

Can Oxygen be Given Safely?

- Yes – if monitored appropriately
 - If CO₂ is monitored in the expired breath or in the blood
- Bi-level respiratory assistance
 - BiPAP v.s. CPAP
 - Assists with the work of breathing
 - Ensures that there is inspiration and expiration



Emergency Care



- Leg Fracture Treatment
 - If your child could walk before the fracture
 - Ask if surgery (internal fixation) may be a better option than casting (will help preserve the muscle and allow walking sooner)
 - Make sure your providers know that this is a priority for you/your child

Fat Embolism Syndrome



Risk with (long bone) fractures

- Medical emergency
- Rapid changes in breathing/breathing rapidly
- Neurologic deterioration –confusion, “not making sense,” “not acting like himself”
- Go **immediately** to the Emergency Department

FES: 2 Major and 1 Minor or 1 major and 4 minor

Major Symptoms	Minor Symptoms
<ul style="list-style-type: none">• Shortness of Breath• Neurologic changes (confusion, headache, coma, seizures)• Petechial rash (develops 24-36 hours after trauma, usually seen in sclera, under arms, chest)	<ul style="list-style-type: none">• Tachycardia (>140's)• Fever• Retinal changes• Jaundice• Decreased PLT• Decreased RBC• Elevated ESR• Fat macroglobulinemia

Fat Embolism Syndrome

- Take the Fat Embolism Syndrome (FES) Care Page with you;
- **Convince the staff that this is an emergency**
- **Assessment**
 - Studies to consider
 - Care for FES
 - » 80% resolve on their own
 - » Symptomatic management
 - » Minimize movements to prevent further fat disruption
- **Best prevention: seat belts in the car AND wheelchair!!**

What is Rhabdomyolysis?

- Result of massive muscle breakdown of skeletal muscle
- Releases myoglobin into the blood
 - Damages the kidney (dark urine)
- Release of potassium into the blood
 - “hyperkalemia”
 - Can result in cardiac arrest



Emergency Care

- Anesthesia

- inhaled anesthesia can cause rhabdomyolysis (NOT malignant hyperthermia)

- All anesthesia should be used with close observation
- **Avoid inhaled anesthesia if at all possible**
- **Absolutely no succinylcholine**

- IV anesthesia is considered safe

- Local anesthesia/nitrous oxide is safe

- Use extreme care with opiate use (avoid if possible)



What Does Cortisol do?

- Hypothalamus-pituitary-adrenal (HPA) Axis



Daily Steroid Dosing

- Daily steroids
 - Cortisol/cortisone coming from elsewhere
 - Temporarily inactivated adrenal glands
- Adrenal crisis
 - Acute adrenal insufficiency
 - Can be life threatening



Emergency Care



- Vomiting for 24 hours
 - Go to ED
 - Risk for Adrenal Crisis
 - Substitute corticosteroid by IV
 - Can figure out IV dose from oral dose
 - 6mg deflazacort = 5 mg prednisone
 - Remind staff that high liver enzymes (AST, ALT) are normal in Duchenne

PJ Nicholoff Steroid Protocol

- Take this with you!
- Deflazacort/prednisone conversion
- Stress doses
- Symptoms of Adrenal Crisis
- Exams/tests to evaluate adrenal function



Thank you!

