DYSTROPHINOPATHY EMERGENCY CARE INFORMATION

FOR FAMILIES

Dystrophinopathy includes Duchenne & Becker muscular dystrophy as well as female carriers of dystrophinopathy. The presentation of these conditions varies, ranging from mild to severe.

NEUROMUSCULAR CENTER/DOCTOR

_____

NEUROMUSCULAR CENTER EMERGENCY NUMBER:

_____

SCAN FOR PJ NICHOLOFF STEROID PROTOCOL

RESPIRATORY CARE

Follow your or your child's pulmonary action plan! If trouble breathing, or oxygen saturation is low, use cough assist or Ambu bag or BiPAP to clear the airway. If breathing does not improve in 5-10 minutes, go to the Emergency Room (ER). Bring all equipment and medications with you to the ER if possible.

LEG FRACTURE TREATMENT

If you or your child has leg pain following a fall, go to Urgent Care or ER to get an X-ray. **If you or your child has difficulty breathing, seems confused, or is less alert after a fall/fracture, this is an emergency! Go immediately to the ER and alert staff that symptoms could be due to Fat Embolism Syndrome (FES) parentprojectmd.org/fes.**

STEROIDS

Remember to tell your doctor if you or your child is on steroids. If severe trauma or unable to take daily corticosteroids for 48 hours, go to the ER and ask that IV corticosteroids are given until pills by mouth are tolerated (6 mg of deflazacort = 5 mg of prednisone = 40mg vamorolone). **Bring the PJ Nicholoff Steroid Protocol** (**parentprojectmd.org/pj**). Stress doses may be needed for moderate/severe stress on the body. Vamorolone cannot be used to stress dose.

GENERAL RECOMMENDATIONS

- Keep immunizations up to date & get influenza (flu) vaccine annually.
- Always wear seat belts in the car AND in wheelchair/scooter/shower chairs.
- Call your neuromuscular team and tell them you are going to the ER/ hospital (do not depend on the ER staff to do this).

ANESTHESIA PRECAUTIONS

If possible, inhaled anesthesia should be avoided. IV anesthesia is considered safe with close monitoring. **Succinylcholine should NEVER be used**. Local anesthesia and nitrous oxide are generally safe for minor dental procedures.

VIEW THIS INFORMATION ONLINE AT PARENTPROJECTMD.ORG/EMERGENCY

Parent Project Muscular Dystrophy — JOIN THE FIGHT. END DUCHENNE.

DYSTROPHINOPATHY EMERGENCY CARE INFORMATION

FOR HEALTHCARE PROVIDERS

Dystrophinopathy includes Duchenne & Becker muscular dystrophy as well as female carriers of dystrophinopathy. The presentation of these conditions varies, ranging from mild to severe.

RESPIRATORY CARE

Risk of respiratory failure. Do not give oxygen without close monitoring of CO2 levels. Breathing may need to be supported (non-invasive ventilation). Use cough assist machine if needed and available.

STEROIDS

Risk of adrenal crisis. Please refer to the **PJ Nicholoff Steroid Protocol** (**parentprojectmd.org/pj**) for stress dosing. Watch for signs of adrenal crisis during times of severe illness, injury, or surgery.

SCAN FOR PJ NICHOLOFF STEROID PROTOCOL

LEG FRACTURE TREATMENT

Risk of pain, loss of ambulation, FES. If ambulatory before leg fracture, surgery is preferred over casting to preserve ambulation (i.e., internal fixation with rapid weight bearing). After any fracture or significant trauma, monitor closely for signs of Fat Embolism Syndrome (FES), which may include:

- Shortness of breath
- Chest pain
- Confusion or altered mental status
- Tachycardia
- Hypotension
- Petechial rash (skin rash)
- Fever

ANESTHESIA PRECAUTIONS

Risk of rhabdomyolysis. Inhaled anesthesia can cause rhabdomyolysis among other serious complications (i.e. cardiac arrest) in patients with dystrophinopathy. When possible, inhaled anesthesia should be avoided. IV anesthesia is considered safe. Use all anesthesia with extreme caution after discussing with the anesthesiologist. **Succinylcholine should NEVER be used.** Local anesthesia and nitrous oxide are generally considered safe for minor dental procedures.

GENERAL RECOMMENDATIONS

- Consider long term steroid therapy when administering live vaccines.
- AST/ALT are normally elevated in individuals with dystrophinopathy and need no further evaluation.
- CK levels are normally elevated in patients with dystrophinopathy. Elevated CK without other symptoms is not an emergency and does not require emergency and/or hospital admission.
- If elevated CK is associated with additional symptoms, such as dark tea-colored urine and/or muscle pain, rhabdomyolysis should be considered.

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