



EMERGENCY INFORMATION FOR PEOPLE WITH DYSTROPHINOPATHY



RESPIRATORY CARE RISK: RESPIRATORY FAILURE

- If trouble breathing, or oxygen saturation low, use cough assist or Ambu bag or BiPAP; if not improved in 5-10 minutes go to ER
- Take your equipment (cough assist, BiPAP, etc.) and medicines with you to the hospital/ER
- In ambulance or ER, medical providers must only give oxygen with close monitoring of CO2 levels; breathing may need to be supported
- Call your Neuromuscular team and tell them you are going to the ER/ hospital. (Do not depend on the staff to do this)

FRACTURES AND FAT EMBOLISM SYNDROME RISKS: PAIN, LOSS OF AMBULATION, AND FAT EMBOLISM

- **Leg Fracture:** If still ambulatory, tell doctor that standing and walking ASAP may preserve ambulation (i.e. internal fixation/surgery with rapid weight bearing)
- 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). To find more information please visit:

parentprojectmd.org/fes

**VIEW THIS INFORMATION ONLINE AT
PARENTPROJECTMD.ORG/EMERGENCY**



RISK FOR ADRENAL INSUFFICIENCY (FOR ALL PEOPLE TAKING STEROIDS)

RISK: ADRENAL CRISIS

- If severe trauma or unable to take daily corticosteroids by mouth for 24 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**
- Stress doses of steroids may be needed for moderate or severe stress on the body. Vamorolone cannot be used to stress dose.

ANESTHESIA PRECAUTIONS

RISK: RHABDOMYOLYSIS (MASSIVE BREAKDOWN OF SKELETAL MUSCLE THAT MAY BE LIFE THREATENING)

- Avoid inhaled anesthesia. IV anesthesia is considered to be safe with close monitoring
- **People with dystrophinopathy should NOT receive succinylcholine**
- Local anesthetics & Nitrous Oxide are safe for minor dental procedures

GENERAL RECOMMENDATIONS AND PRECAUTIONS

- Keep immunizations up to date and get influenza (flu) vaccine annually
- People taking daily, long term steroids should ask their doctor if it safe to get live vaccines
- Always wear seat belts! This includes in the car AND in chairs/wheelchairs/scooters/shower chairs
- Remind clinicians that some "liver" tests (AST/ALT) are normally elevated in patients with dystrophinopathy. That is because AST/ALT also come from muscle, and require no further testing
- Remind clinicians that CK (creatinine kinase) is normally elevated in patients with dystrophinopathy. Elevated CK without other symptoms is not an emergency and does not require emergency care 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.