IN AN EMERGENCY REMEMBER TO

THINK

Parent Project Muscular Dystrophy
JOIN THE FIGHT. END DUCHENNE.

T TAKE YOUR EQUIPMENT WITH YOU TO THE HOSPITAL

- Bipap
- Cough Assist

H HAND YOUR EMERGENCY CARE CARD TO EMERGENCY MEDICAL TEAM TO REVIEW

- Emergency Care Card
- Highlight signs of FES if a recent fall or trauma has occurred

I INFORM MEDICAL STAFF OF IMPORTANT OXYGEN PRECAUTIONS

- Only use with pressure support or with close CO2 monitoring

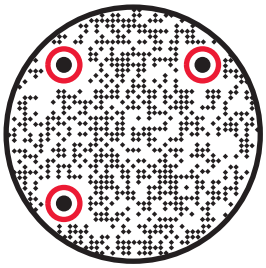
N NOTIFY YOUR NEUROMUSCULAR TEAM

- Reference your emergency contact information

K KEEP IMPORTANT DOCUMENTS WITH YOU

- Copy of last neuromuscular note
- Medication List
- Copy of the PJ Nicholoff Steroid Protocol
- “About Me” information sheet
- Legal paperwork

SCAN ME FOR RESOURCES



PJ NICHOLOFF STEROID PROTOCOL

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ABOUT THIS DOCUMENT

This document is meant for healthcare providers. For further explanation of steroids and adrenal crisis, please see the PPMD Steroid Care Page at **parentprojectmd.org/steroids**.

BACKGROUND/ASSESSMENT

Normal basal secretion of cortisol from the adrenal gland is approximately 5-7 mg/m2/day or 8 -10 mg/day for adults. This amount increases during minor illnesses or surgery to approximately 50 mg/day (5x normal physiologic secretion). These small increases with uncomplicated surgery return to baseline in 24 hours. Procedures producing greater surgical stress, have been shown to increase cortisol responses to 75-150 mg/day (10x normal physiologic secretion), which return to baseline in about 5 days.

Corticosteroids are prescribed for multiple diagnoses to a wide variety of patients. Long term administration of corticosteroids may lead to suppression of the hypothalamic-pituitary-adrenal (HPA) axis. Rapid reduction or abrupt withdrawal of corticosteroid therapy that has been prolonged or at high doses can cause secondary adrenal insufficiency (suppression of the HPA axis), and steroid withdrawal or deprivation syndrome. Recovery from suppression of the HPA axis after discontinuing corticosteroids can be prolonged (possibly 6 to 12 months) and may vary based on doses, dosing schedules and duration of corticosteroid therapy. Since there is a great deal of individual variability in susceptibility to suppression of the HPA axis after chronic use of exogenous corticosteroids, it is not possible to predict with confidence which patients will be affected. Current practice is to administer supplemental (stress) doses of corticosteroids to patients with suspected suppression of the HPA axis in the perioperative period and during acute illness to prevent acute adrenal insufficiency, or adrenal crisis.

DEFINING HPA SUPPRESSED PATIENTS:

Recommendations differ slightly in defining a suppressed patient, but general guidelines are below (Table 1):

Prednisone dose Equivalents/day - Adults	Prednisone Dose Equivalents/Day - Pediatric	Suppression of HPA axis?
5mg/day or less	3 mg/m2/day or less	Usually not suppressed.
5 – 20 mg/day	3-12 mg/m2/day	Possibly suppressed. ACTH stimulation test recommended or give supplemental dose.
20 mg/day for >10 days or more	12 mg/m2/day of prednisone for > 10 days or more	Suppressed. Give supplemental dose.

Patients receiving disease appropriate corticosteroid doses (at least 10 times above the physiologic cortisol dose) generally do not need stress doses if usual daily dose is continued. Patients who are on maintenance physiologic dose of hydrocortisone for primary disease of the HPA axis do require supplemental therapy.

Consultation with endocrinology is recommended for questions or concerns.

Recommendations for supplemental doses are generally divided by severity of stress the patient may experience (medical or surgical). Supplemental steroid doses are then based on degree of stress.

CORTICOSTEROID STRESS DOSES:

Table 2

Medical/Surgical Stress	Corticosteroid Dosage DOS*	Postoperative Taper Regimen
Minor (local anesthesia, < 1 hour) (e.g. inguinal hernia, single tooth extraction, colonoscopy), mild febrile illness, mild, nausea/vomiting, mild diarrhea)	25mg or 30-50 mg/m2 po (if able to take po) or IV hydrocortisone (HC) or equivalent	None Resume maintenance physiologic dose of hydrocortisone when illness, pain or fever subsides
Moderate (e.g. multiple teeth extraction, fracture, pneumonia)	50mg or 50-75 mg/m2 IV hydrocortisone or equivalent	25 mg Q 8 or 50-75 mg/m2/day ÷ q 6 hours X 24 hour. Taper to baseline over 1-2 days.
Major (e.g. Septic shock, multiple trauma/fractures or severe burns, severe systemic infections, major surgery, pancreatitis, orthopedic surgery including open reduction, spinal fusion, etc.)	100mg or 100 mg/m2/dose IV hydrocortisone or equivalent	50 mg IV Q 8 or 100 mg/m2/day ÷ q 6 hours X 24-48hours. Taper to baseline over 1-3 days (continue stress dose if the physical stress (fever or pain) continues).

DOS - Day of surgerys

Patients using high dose twice-weekly corticosteroid-dosing schedule:

- If patients using a twice-weekly dosing schedule are unable to take corticosteroids by mouth during a time when they should be taking corticosteroids (due to nausea, vomiting, diarrhea, etc.), patients should take stress doses intravenously as indicated above.
- If patients using a twice weekly dosing schedule are undergoing/experiencing a moderate or major medical/surgical stressor in their life, cortisol level should be drawn and it is recommended that they follow the stress dosing recommendations in the above table.
- No literature exists for these recommendations. The recommendations are based on expert opinion and practice.

RECOMMENDATION FOR CORTICOSTEROID THERAPY WITHDRAW:

Below is one recommendation for tapering chronic corticosteroids (generally managed in an outpatient setting):

- Start on a Monday, giving 20-25% lower corticosteroid dose for 2 weeks (or longer)
- If multiple doses are taken, start first to reduce multiple daily doses to a single morning dose
  - Cut the dose 20-25% again for 2 weeks (or longer); continue this schedule
  - Continue until dose is near physiologic dose (3mg/m2/day of prednisone or 3.6mg/m2/day of Deflazacort)
- When near physiologic dose, substitute corticosteroids with short acting form of corticosteroid or hydrocortisone (12 mg/m2/day of hydrocortisone)
- This will also enable the patient to have a supply of hydrocortisone to be used for stress doses if needed in times of stress after coming off steroids
  - Continue to taper off by 20-25% each week (or longer)
  - Give every other day for 2 weeks (or longer)
  - Stop
  - WATCH VERY CAREFULLY FOR SIGNS OF ADRENAL CRISIS (see below)
- Alert parents to signs/symptoms of adrenal crisis
- If patients have symptoms of adrenal insufficiency during the taper, the steroid dose prior to the taper should be maintained for longer

IF THE PATIENT HAS A SERIOUS ILLNESS/INJURY DURING THE TAPER, THEY MAY NEED A "STRESS DOSE" OF CORTICOSTEROIDS:

- Encourage parents to continue to report any serious events until 1 year post-taper
  - The stress doses of hydrocortisone is 30-50 mg/m2/day, or higher, for major stress (see Table 2)
  - Patients need to go to the emergency room if having signs or symptoms of adrenal crisis. Serum electrolytes with blood glucose and cortisol level should be obtained.
  - Patients should see a pediatric endocrinologist for evaluation of HPA axis during the process of corticosteroid therapy withdrawal.

Patients using high dose twice-weekly corticosteroid-dosing schedule:

- It is recommended that patients electing to discontinue the use of twice weekly corticosteroids, do so under the guidance of a neuromuscular provider and/or endocrinologist.
- No literature exists for these recommendations. The recommendations are based on expert opinion and practice

Testing HPA axis:

- After reaching half the physiological dose (5-6 mg/m2/day of hydrocortisone or 1-1.5 mg/m2/day of prednisone), morning serum cortisol and ACTH should be assayed monthly (may do less frequently), until they reach normal levels.
- When baseline morning serum ACTH and cortisol are normal, discontinue the corticosteroid and carry out the rapid ACTH stimulation test monthly until post-stimulation cortisol response is normal (post-stimulus level > 20 mcg/dL). When this point is reached, it can be considered that the HPA axis has recovered

Modification of above protocol:

- Omit monthly AM cortisol and ACTH and perform an ACTH stimulation test in 3 months after discontinuation of corticosteroids
- During this time (3 months before getting ACTH stimulation test), patients will need to take stress dose at the time of stress
- If ACTH stimulation test result is abnormal (peak cortisol <20), patients will need to continue taking stress doses of hydrocortisone at the time of stress. (Patients Repeat ACTH stimulation test again in 1-2 months later and families would need to have teaching on this with an endocrine nurse.)

Alternatively, when laboratory tests cannot be carried out:

- Patients who have used corticosteroids for prolonged periods can be considered as having suppression of the HPA axis up to 1 year after discontinuation of corticosteroid therapy and therefore need hydrocortisone stress dose coverage during the time of stress

Risk factors for adrenal crisis include:

- Dehydration
- Infection and other physical stress
- Injury to the adrenal or pituitary gland
- Stopping treatment too suddenly with glucocorticoid medications such as prednisone hydrocortisone
- Surgery
- Trauma

Symptoms of adrenal crisis can include any of the following:

- Abdominal pain
- Shock
- Confusion or coma
- Dehydration
- Dizziness or light-headedness
- Fatigue
- Flank pain
- Headache
- High fever
- Loss of appetite
- Loss of consciousness
- Low blood pressure
- Nausea
- Profound weakness
- Rapid heart rate
- Rapid respiratory rate (see tachypnea)
- Slow, sluggish movement
- Unusual and excessive sweating on face or palms
- Vomiting

Exams and Tests

Tests that may be ordered to help diagnose acute adrenal crisis include:

- ACTH (cosyntropin) stimulation test
- Cortisol level
- Blood sugar
- Serum potassium
- Serum sodium
- Serum pH

Corticosteroid Conversion Table

Table 3

Medication	Equivalent doses
Cortisone	25 mg
Hydrocortisone	20 mg
Deflazacort	6 mg
Prednisone	5 mg
Methyl prednisone	4 mg
Triamcinolone	4 mg
Betamethasone	0.75 mg
Dexamethasone	0.75 mg

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Acknowledgements

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- Dr. Anne M. Connolly, St. Louis Children's Hospital, St. Louis, MO

In honor of the late Philip James "PJ" Nicholoff, for his contribution to the global Duchenne community.

DUCHENNE MUSCULAR DYSTROPHY EMERGENCY CARE INFORMATION FOR FAMILIES

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RESPIRATORY CARE

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

LEG FRACTURE TREATMENT

If your child has leg pain following a fall, go to Urgent Care or ER to get an X-ray. **If 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).**

STEROIDS

Remember to tell your doctor if 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 equals 5 mg of Prednisone).** Bring the PJ Nicholoff Steroid Protocol (parentprojectmd.org/pj). Stress doses may be needed for moderate/severe stress on the body.

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.

GENERAL RECOMMENDATIONS

- Keep immunization up to date & get influenza (flu) vaccine annually.
- Always wear seat belts in the car AND in chairs/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).

NEUROMUSCULAR CENTER/DOCTOR: _____

NEUROMUSCULAR CENTER EMERGENCY NUMBER: _____

DUCHENNE MUSCULAR DYSTROPHY EMERGENCY CARE INFORMATION FOR HEALTHCARE PROVIDERS

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.

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). Following a fracture or body trauma, **watch for signs of Fat Embolism Syndrome (FES)** including fast breathing and/or confusion.

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.

ANESTHESIA PRECAUTIONS

Risk of rhabdomyolysis. Inhaled anesthesia can cause rhabdomyolysis among other serious complications (i.e., cardiac arrest) in patients with Duchenne. 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 safe for minor dental procedures.

GENERAL RECOMMENDATIONS

- Consider long term steroid therapy when administering live vaccinations.
- AST/ALT are normally elevated in patients with Duchenne and need no further evaluation.

IMPORTANT CONTACT INFORMATION

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If you find yourself at the hospital with a medical emergency, be sure to be involved in the plan of care. In addition to sharing your PPMD emergency card with clinicians involved in your care, it is also important to make sure a physician with experience in Duchenne is involved in your or your child's care in the hospital.

We have compiled a list of contact numbers from each of the PPMD Certified Duchenne Care Centers (CDCCs) below to ensure a Duchenne specialist can be involved in your child's plan of care, and make sure this happens:

1. Call your neuromuscular team/center.
2. Call the hospital where you receive neuromuscular care if it is after hours.
3. Physician to physician calls.

If you have questions about planning ahead in case of emergencies, please don't hesitate to contact PPMD's Care Team at careteam@parentprojectmd.org.

1) CALL YOUR NEUROMUSCULAR TEAM/CENTER

It is critical to notify your neuromuscular care team if you are in the hospital with an illness or injury. This is especially important if you are being seen at a hospital that is not affiliated with your neuromuscular care.

It is always important to notify your care team so they can work with the clinicians taking care of you or your child to ensure you receive the best care possible. Do not depend on the hospital staff to communicate with your neuromuscular care team; it is always best to personally confirm your neuromuscular care team is involved.

2) CALL THE HOSPITAL WHERE YOU RECEIVE NEUROMUSCULAR CARE IF IT IS AFTER HOURS

We all know emergencies can happen in the middle of the night or on a weekend when it may be more difficult to contact your neuromuscular center. It is best to discuss an emergency plan with your neuromuscular specialist and ask for a number to call outside of regular office hours.

If you do not have a direct number to a specialist on call, your next option is to call the main hospital's operator and ask for the specialist on call (i.e. Neurologist, Pulmonologist, Cardiologist, etc). From here, they should be able to connect you with the appropriate specialist and help drive your care.

3) PHYSICIAN TO PHYSICIAN CALLS

You have the right to ask for a second opinion. It can be intimidating to question a physician about their plan of care. However, even with the best of intentions, they may not be treating you or your child in a way that is appropriate or safe for someone living with Duchenne.

Ask the physician taking care of you or your child to connect with your neuromuscular team (if you are cared for at a CDCC) or a specialist from one of the CDCCs (if you are not seen at one of these locations) to discuss your or your child's case. Each CDCC is located at a hospital that has a physician consult line, which you can give to the doctor taking care of your or your child.

CDCC CONTACT INFORMATION

Center	Neuromuscular Clinic	After Hours	Physician to Physician
Akron Children's Hospital	330-543-6014	330-543-1000	330-543-8050
American Family Children's Hospital	608-890-6500/ 608-263-6420	608-263-6400	608-263-3260
Ann and Robert H. Lurie Children's Hospital	312-227-3550	312-227-4000	312-227-3560
Arkansas Children's Hospital	501-364-1850	501-441-3453	501-441-3453
Billings Clinic	406-238-5770	406-238-5800	406-238-5107
Boston Children's	617-355-8235		617-355-6363
Children's Hospital Colorado	720-777-2806	720-777-1234	720-777-3999
Children's Hospital of the King's Daughters	757-668-9729	757-668-7000	757-668-9999
Children's Hospital of Richmond VCU	804-628-5804	804-828-0951	804-828-0442
Children's Hospital Wisconsin	414-266-3464	414-266-2000	414-266-2460
Children's Medical Center Dallas	214-456-2768, opt 2, opt 5	214-456-7000 and ask to speak with ON-CALL NEUROLOGIST	214-456-7000 and ask to speak with ON-CALL NEUROLOGIST
Children's Mercy Hospital Kansas City	816-302-3387	816-234-3000	1-800-466-3729
Children's National Hospital	202-476-6193	202-476-5000	202-476-2120
Cincinnati Children's Hospital Medical Center	513-803-3000, option 2	513-636-4200	513-636-4010
Duke University	919-613-6832	919-684-8111 (ask for MD to be paged)	919-416-3853
Helen DeVos Children's Hospital	616-267-2500	616-391-9000	616-267-2500
Kennedy Krieger Institute	443-923-9525	443-923-9200	443- 923-9403

Center	Neuromuscular Clinic	After Hours	Physician to Physician
Lucile Packard Children's Hospital Stanford	650-723-0993	650-497-800	800-995-5724
Nationwide Children's Hospital	614-722-2203	614-722-2000	614-355-0221
Nemours AI Dupont	302-651-5930	302-651-4000	302-651-4000
Penn State Health Children's Hospital	717-531-0003 ext 282670		800-233-4082
Phoenix Children's Hospital	602-933-0970	602-933-0970	602-933-0970
Riley Children's	317-948-7450	317-948-7450	800-622-4989
Seattle Children's	206-987-6678 option 5	206-987-2000	206-987-7777, option 4
St. Louis Children's	314-362-6981	314-454-6000	314-362-6981
Stony Brook	631-216-8195	631-216-8195	631-444-2725
UC Davis	916-734-7041	916-734-2011	800-482-3284, option 2
UCLA	310-794-1195	415-476-1000	310-405-9824
UCSF Benioff Children's Hospital	415-514-8848	415-476-1000	877-822-4453
University of Iowa	319-353-6200	319-356-1616	319-356-7726
University of Louisville/ Norton Children's Hospital	502-588-9590	502-588-3650	502-588-3650
University of Rochester	585-275-2559	585-275-2559	585-275-2559
University of Utah/Primary Children's Hospital	801-885-7575/ 801-213-3599	801-662-1000	801-213-7789
University of Virginia	434-924-8184	434-924-0000—page Developmental Pediatrics on-call provider	434-924-5491
UPMC Children's Hospital of Pittsburgh	412-692-5325	615-936-1000	615-936-4444
Vanderbilt	615-875-8500	615-936-1000	615-936-4444
Yale New Haven Children's Hospital	203-688-4221	203-688-4242	888-964-4233
Other:			

*Please be sure to confirm contact information with your center