

Emergency Instructions

Treatment for Congenital Adrenal Hyperplasia in times of stress

Includes Information on:

- **Stress Dosing**
- **Hydrocortisone Injection**
- **Emergency Room Instructions**

Your or your child's body does not make enough of certain essential hormones: cortisol, and in salt wasting CAH, aldosterone. Cortisol, which is produced by the adrenal glands, has many purposes in the body such as maintaining energy supply, fluid, electrolyte balance, blood pressure, normal blood sugar levels, and controlling the body's reaction to physical stress. Aldosterone is used by the kidneys to maintain a normal blood sodium and fluid balance (salt and water). When cortisol and aldosterone are not produced by the body they must be replaced by medication.

Extra hydrocortisone must be given during times of extreme physical stress such as fever, vomiting and diarrhea, surgery, and traumatic injuries (e.g., broken bones and concussions). The extra hydrocortisone is called a "stress dose." The Florinef dose does not change. Make sure you discuss stress dosing with your or your child's physician and you know how to proceed in the event of illness.

If ill, call physician to alert him/her of your or your child's condition. Typically, stress dosing is required when . . .

***FEVER IS GREATER THAN 100.5°F** : DOUBLE the hydro-cortisone dose for the entire day

***FEVER IS GREATER THAN 102°F** : TRIPLE the hydrocortisone dose for the entire day

***VOMITING**: Triple dose with vomiting with or without a fever. If you vomit less than 30 minutes after taking the hydrocortisone stress dose, the medication likely was not absorbed and the dose should be repeated. Wait 10-15 minutes after you/your child vomit(s) and repeat triple stress dose of hydrocortisone. If you/your child vomit(s) again, give the injectable hydrocortisone (brand name Solu-Cortef® in the U.S.) and contact your physician. **DO NOT DELAY in giving the injectable hydrocortisone.**

***DIARRHEA**: Injection may also be needed in the event of diarrhea due to loss of fluids. If diarrhea, no fever and feeling fine, no need to stress dose. If not feeling well, double dose of hydrocortisone recommended.

*Try small amounts of clear liquids that contain SUGAR (not artificial sweetener) frequently, at least 1 ounce every 15 minutes.

Signs of acute adrenal crisis from cortisol:

- Headache
- Abdominal pain
- Pale skin
- Dehydration
- Nausea
- Confusion
- Listlessness
- Dizziness

If these occur and continue after oral stress dosing, call your or your child's physician and go to the nearest emergency room immediately.

Again, do NOT wait to give the injectable hydrocortisone. It should be given BEFORE a trip to the emergency room or activating 911 if those actions become necessary.

Remember, stress dose with:

Fever of 100.5°F or higher • Vomiting • Diarrhea
Physical trauma (broken bone, concussion, etc.)

EXAMPLE STRESS DOSING:

Normal dose: 1 tab + 1/2 tab + 1 tab (total of 2.5 pills)

Double dose: 5 total tablets (divide into 1.5 tablets every 8 hours)

Triple dose: 7.5 total tablets (divide into 2.5 tablets every 8 hours)

HOW TO GIVE AN INJECTION OF HYDROCORTISONE INSTRUCTIONS

1. STAY CALM. Wash your hands and gather equipment: needle, syringe, alcohol pad, and vial of hydrocortisone (Solu-Cortef® Act-O-Vial).



Steps 2-3



Steps 4-5

2. Mix the medication by pushing down on top of the vial to release the cork into the vial.

3. Shake the vial to mix medicine, take off the top of the vial, and wipe down the rubber stopper with alcohol.



Steps 6-7



Steps 8-10

4. Take the cap off the syringe needle and insert into the vial through the rubber stopper.

5. Draw up the medication and replace the needle cap.

6. Select the site for the intramuscular injection, typically the outer portion in the middle of the thigh.

7. Use the alcohol to clean the skin at the injection site.



Step 11



Step 12

8. Take off the cap of the needle and hold the syringe like a dart.

9. Using your thumb and first two fingers, spread the skin by pushing down lightly.

10. Dart the needle into the thigh, going at a 90° angle.

11. Hold the syringe in place and pull back the plunger to make sure you don't see blood, (seeing blood means you are in a blood vessel, but this is rare). If you do see blood, withdraw syringe and discard*. Prepare another syringe with medication and inject in a slightly different site. (*However, if this is the only dose you have, continue with the same syringe, injecting in a slightly different site).

12. Inject medicine then place tissue or cotton ball near the needle. Pull the needle out quickly.

13. Place the needle and syringe in a hard, unbreakable container.

14. Call doctor, 911, or go to hospital, (if necessary).

EMERGENCY ROOM INSTRUCTIONS We recommend filling out this form with your/your child's name, then giving it to your/your child's physician (whichever physician you would like contacted in case of emergency). Have the doctor read over, complete & sign. You and your child's caregiver should always carry a copy of these signed instructions.

I/my child, _____, have/has a rare, inherited, genetic disorder called Congenital Adrenal Hyperplasia (CAH). I am/my child is adrenally insufficient and steroid dependent. I/my child must be seen by a physician IMMEDIATELY because life threatening electrolyte disturbances/adrenal crises are possible with febrile illnesses, fluid depletion from vomiting and diarrhea, surgery, and serious injuries.

Time in a waiting area or triage situation is not appropriate.

Signs of adrenal crisis include, but are not limited to weakness, dizziness, nausea and vomiting, hypotension, hypoglycemia, pallor, and lethargy.

Treatment should include:

*IV fluids-D5 normal saline at 20cc/kg for at least one hour then continuous fluid replacement for dehydration and hypotension.

*STAT basic metabolic panel (sodium, potassium, chloride, carbon dioxide, glucose, BUN, creatinine, and calcium)

*Initial hydrocortisone IV bolus can be administered IM if IV access an issue

- 25mg for children under age 3
- 50mg for children aged 3-10
- 100mg for children older than 10 years or weighing more than 40kg
- 100mg for teens and adults

*Hydrocortisone as a continuous drip (if necessary) or in 4 divided doses IV bolus

- 50mg/day for ages 0-37
- 75mg/day for ages 3-10
- 100mg/day for children older than 10 years or weighing more than 40kg
- 100mg/day for teens and adults

NOTE FROM PHYSICIAN: Please follow the above treatment instructions and contact me as soon as possible. (See contact info below). Patient's health issues include:

Thank you. Please call if additional information is needed.

_____ (Physician signature)

Physician Contact Information

Endocrinologist: _____

Phone Number _____ **Fax Number** _____

Primary Care Provider/Pediatrician: _____

Phone Number _____ **Fax Number** _____

These are the medications I/my child take(s) daily: _____

These are the medications I have/my child has **taken today** (includes any stress dosing), including approximate time(s) medication(s) was/were taken: _____

