

## Physician Contact Information

Endocrinologist:

\_\_\_\_\_ M.D.

Phone Number \_\_\_\_\_

Fax Number \_\_\_\_\_

Pager \_\_\_\_\_

Primary care provider/Pediatrician:

\_\_\_\_\_ M.D.

Phone Number \_\_\_\_\_

Fax Number \_\_\_\_\_

**These are the medicines I/my child take(s) on a daily basis:**

\_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_

**These are the medicines I have/my child has taken today (includes any stress dosing for the day):**

\_\_\_\_\_  
\_\_\_\_\_  
\_\_\_\_\_

## EMERGENCY ROOM 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 crisis 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 impending 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 20 cc/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

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

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

- 50mg/day for 0-3 years of age
- 75 mg/day for 3-10 years of age
- 100 mg/day for children greater than 10 years of age or weighing more than 40 kg
- 100 mg/day for teens and adults

**Please contact my endocrinologist as soon as possible. (see panel to left) health issues:**

\_\_\_\_\_  
\_\_\_\_\_

**Thank you and please call with any questions.** \_\_\_\_\_ **M.D.**

## 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 would require these actions:

**\*FEVER GREATER THAN 100.5: DOUBLE the hydrocortisone dose for the entire day**

**\*FEVER GREATER THAN 102: TRIPLE the hydrocortisone dose for the entire day**

**\*VOMITING:** Triple dose with vomiting whether or not there is 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/r child vomit(s) and repeat triple stress dose of hydrocortisone. *If you/r child vomit(s) again, give the injectable hydrocortisone (brand name Solu-Cortef® in the US) 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 deficiency: **HOW TO GIVE AN INJECTION OF HYDROCORTISONE**

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

If these occur and continue after stress oral 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 greater
- Vomiting
- Diarrhea
- Physical trauma (broken bone, concussion, etc.).

**EXAMPLE OF ORAL STRESS DOSING**

	Normal dose	Double dose	Triple dose
Morning	1 pill	2 pills	3 pills
Afternoon	1/2 pill	1pill	1 <sup>1</sup> / <sub>2</sub> pills
Bedtime	1/2 pill	1 pill	1 <sup>1</sup> / <sub>2</sub> pills

1. STAY CALM. Wash your hands and gather equipment: needle, syringe, alcohol pad, and vial of hydrocortisone (Solu-Cortef® Act-O-Vial).
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.
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.
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 and push down lightly.
10. Dart the needle into the thigh, going at a 90 degree angle.
11. Hold the syringe in place and pull back the plunger to make sure you don't see blood (which would mean you are in a blood vessel\*. If you do (which would be rare), 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.



**Steps 2-3**

Mix the medication and shake the vial.



**Steps 4-5**

Insert the needle into the vial and draw up the medication.



**Steps 6-7**

Select the site for injection and use the alcohol to clean the skin.



**Steps 8-10**

Hold the syringe like a dart, spread the skin pushing down slightly, and dart the needle into the thigh at a 90 degree angle.



**Step 11**

Pull back the plunger to check for blood.



**Step 12**

Inject medicine then place tissue/cotton ball near the needle and pull needle out quickly.