Emergency Instructions

Caring for your child with Congenital Adrenal Hyperplasia in times of stress

Includes Information On:

Stress Dosing

Hydrocortisone Injection

Emergency Room Instructions

www.caresfoundation.org  info@caresfoundation.org  1-866-227-3737
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, maintaining fluid and electrolyte balance, maintaining blood pressure, maintaining 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 such as broken bones and concussions. The Florinef dose does not change. The extra hydrocortisone is called a “stress dose.” Make sure you discuss stress dosing with your child’s physician and you know how he/she would like you to proceed in the event of an illness.

If your child becomes ill, call the child’s physician to alert him/her of the 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 or DIARRHEA: If your child vomits, wait 30 minutes and repeat the dose (double or triple according to child’s needs). If your child still vomits that oral dose, you need to give the injectable hydrocortisone (brand name Solu-Cortef in the US) and contact the child’s physician. DO NOT DELAY in giving the injectable hydrocortisone. Offer small amounts of clear liquids that contain SUGAR frequently, at least 1 ounce every 15 minutes. Injection may also be needed in the event of diarrhea due to loss of fluids. Remember that children tend to get sicker at night, so if in doubt, stress dose before bedtime.

Watch for signs of acute adrenal crisis from cortisol deficiency:
- Headache
- Nausea
- Abdominal pain
- Confusion
- Pale skin
- Listlessness
- Dehydration
- Dizziness

If these occur and continue after stress dosing, call the child’s physician and take child 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 when:
- Your child is sick with a fever of 100.5°F or greater.
- Your child is vomiting.
- Your child has diarrhea.
- Your child has experienced physical trauma (broken bone, concussion, etc.).

**EXAMPLE OF ORAL STRESS DOSING**

<table>
<thead>
<tr>
<th></th>
<th>Normal dose</th>
<th>Double dose</th>
<th>Triple dose</th>
</tr>
</thead>
<tbody>
<tr>
<td>Morning</td>
<td>1 pill</td>
<td>2 pills</td>
<td>3 pills</td>
</tr>
<tr>
<td>Afternoon</td>
<td>1/2 pill</td>
<td>1 pill</td>
<td>1½ pills</td>
</tr>
<tr>
<td>Bedtime</td>
<td>1/2 pill</td>
<td>1 pill</td>
<td>1½ pills</td>
</tr>
</tbody>
</table>
HOW TO GIVE AN INJECTION OF HYDROCORTISONE

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. My child's dose is _____mg of hydrocortisone which is _____ml of Solu-Cortef.

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, meaning 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, continue with the same syringe, injecting in a slightly different site).

12. After injecting the medicine, place tissue or cotton ball near the needle and 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
Place tissue or cotton ball near the needle and pull needle out quickly.

*Pictures reprinted with permission from the National Institutes of Health.*
EMERGENCY ROOM INSTRUCTIONS

This child has a rare, inherited, genetic disorder called Congenital Adrenal Hyperplasia (CAH). Basically, he/she is adrenal insufficient and steroid dependent. He/she must be seen by a physician IMMEDIATELY because life threatening electrolyte disturbances/adrenal crisis are highly 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 continue fluid replacement
*STAT basic metabolic panel (sodium, potassium, chloride, carbon dioxide, anion gap, glucose, BUN, creatinine, and calcium)
*Initial Solu-Cortef IV bolus under these guidelines—can be administered IM if IV access an issue
-25 mg for children under 3 years of age
-50 mg for children 3-10 years of age
-100mg for children older than 10 years of age
*Solu-Cortef as a continuous drip (if necessary) or in 4 divided doses
-50 mg/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

Please contact the Pediatric Endocrinologist as soon as possible. (see panel to left)

These are the child’s other health issues:


Thank you and please call with any questions.

_________________________________________ M.D.