**Adrenal Insufficiency**

**What is Adrenal Insufficiency (AI)?**

Adrenal Insufficiency occurs when the adrenal glands do not make enough of certain hormones, such as cortisol. Cortisol is the hormone often referred to as the “stress hormone” or the “fight or flight hormone.” In some forms of adrenal insufficiency, the adrenal glands do not make the hormone aldosterone. Adrenal insufficiency is treated by replacing the hormones the body doesn’t make. **Children with adrenal insufficiency can have normal, active lives.** Adrenal insufficiency may be permanent or temporary. *Without proper treatment, adrenal insufficiency can be life-threatening.*

**What does cortisol do?**

Cortisol has many functions, including:

* Keeps the blood sugar in a normal range
* Aids in energy production and metabolism
* Maintains a normal blood pressure
* Helps your body respond to stress
* Enables the body to recover from illness, injury, or surgery. *During these times, it is necessary for the body to make more cortisol.*

**What does aldosterone do?**

Aldosterone helps keep the right amount of sodium, potassium, and water in the body. One function of this balance is maintaining a normal blood pressure. Sodium and potassium are important electrolytes that help maintain normal nerve and muscle function and a regular heartbeat.

**What causes Adrenal Insufficiency?**

**Primary Adrenal Insufficiency**: Occurs when the adrenal glands are damaged and unable to make enough cortisol and aldosterone. *This form of adrenal insufficiency is permanent.*

* + Addison’s Disease – adrenal glands are damaged due to an autoimmune process
  + Damage or removal of both adrenal gland
  + Defect of adrenal glands from birth
  + Congenital Adrenal Hyperplasia (CAH) – adrenal gland dysfunction with sex organ involvement

**Secondary Adrenal Insufficiency:** Occurs when the pituitary gland doesn’t make enough ACTH to tell the adrenals to make cortisol. *This form of adrenal insufficiency may be permanent or temporary.*

* + Long-term steroid treatment
  + Defect of pituitary gland from birth
  + Tumors, radiation, and/or pituitary surgery
  + Traumatic head injury

**What are signs and symptoms of Adrenal Insufficiency?**

Symptoms of Adrenal Insufficiency (AI) are often vague and can be attributed to other health problems or medication side effects. For this reason, AI can be difficult to diagnosis and diagnosis may be delayed.

* Weakness
* Fatigue
* Weight loss
* Decreased appetite
* Stomach pain
* Nausea or vomiting
* Dizziness
* Fainting
* Leg or back pain
* Joint or muscle aches
* Low blood pressure
* Low blood sugar
* Irritability
* Depression

**How is Adrenal Insufficiency treated?**

Adrenal insufficiency is treated by replacing the hormones that the body is unable to make.

**Cortisol** **must be replaced in all types of Adrenal Insufficiency.** Cortisol is replaced with a steroid, like hydrocortisone. Other medications, like prednisone may also be used.

The goal is to replace the amount the body should be making. This is called **maintenance dosing**. *This amount changes as children grow, so regular follow up and dose adjustments are critical.* Doses are usually given two (every 12 hours) or three times a day (every 6-8 hours).***It is important to give this medication every day to avoid Adrenal Crisis.***

If there are other hormone deficiencies, such as aldosterone, these will need to be replaced as well. Aldosterone is replaced with a mineralocorticoid, like fludrocortisone in Primary Adrenal Insufficiency.

In salt-wasting Congenital Adrenal Hyperplasia, salt may need to be added to the diet for infants who do not get enough salt from breast milk or formula. These children may require other treatment as well, particularly if they have ambiguous genitalia.

**What is “Stress dosing” or sick day management?**

During times of illness or injury the body needs more cortisol to stay healthy. A person with Adrenal Insufficiency cannot make cortisol, so higher amounts of steroid medication need to be given. This is typically triple the amount of daily doses. ***Refer to your child’s Adrenal Insufficiency Action plan for dosing details.***

Stress dosing is needed if:

* Fever of 100.4 F or higher
* Illness that keeps your child home from school (not for just a minor cough/cold)
* Vomited once
* Diarrhea once
* Trauma, such as broken bone or car accident
* Severe grief

Stress dosing needs to be continued until child is back to baseline or is fever free for 24 hours. Then resume normal maintenance dosing. ***Please contact your provider if having to stress dose for more than 7 days.***

You should also have your child drink plenty of sugar and salt-continuing fluids to prevent dehydration, low blood sugar, and/or low sodium.

**What if my child is too sick to take medication by mouth?**

If your child cannot take medication by mouth or their body is not able to use their medication because of vomiting or diarrhea, then an injection of their steroid medication needs to be given. **This is urgent!**

The injection needs to be given for:

* Repeated vomiting – more than once
* Watery diarrhea – 2 times or mor in a 4-hour period
* Unable to take oral medication
* Major trauma
* Seizure
* Unconsciousness

***\*Adrenal Crisis can occur if this medication is not given. Do not hesitate to give this injection - it is safe and life-saving! \****

**After giving the injection, your child needs to be seen in the emergency room to ensure further care is not needed. Call 911 or go to your nearest ER. Take your emergency letter with you.**

**What are signs of Adrenal Crisis?**

These symptoms may look a lot like those at diagnosis and can include any number of the following:

* Stomach pain
* Nausea or vomiting
* Diarrhea
* Weight loss
* Loss of appetite
* Fatigue or lethargy
* Weakness
* Headache
* Confusion
* Dizziness
* Fainting
* Low blood pressure
* Joint or muscle pain
* Shakiness or restlessness
* Pale skin
* Cool
* Sweaty
* Irritability
* Signs of dehydration (dry mouth, sunken eyes or dark circles under the eyes, not peeing as much as normal)
* Fast heart rate

**When in doubt, give injection and seek emergency care!**

**What if my child is having surgery or a procedure?**

Higher doses of medications are needed during and after surgery or a procedure (including dental). Ensure the surgeon or dentist is aware of the Adrenal Insufficiency diagnosis and medications. They should contact the Endocrinology Clinic for orders before surgery/procedure.

**Managing Adrenal Insufficiency**

* **Remember, your child can still lead an active life with Adrenal Insufficiency!**
* Always carry your child’s medications and Adrenal Insufficiency Action Plan with them.
* They should wear a medical identification that states, “Adrenal Insufficiency”.
* Make sure all caregivers – including school and daycare - have medication and stress dosing instructions.
* Your child should be seen regularly in the Endocrinology Clinic.

**Steps to Administer Solu-Cortef Injection**

You may also view a step-by-step video on our website: [www.childrensomaha.org/department/endocrinology-diabetes/](http://www.childrensomaha.org/department/endocrinology-diabetes/)

|  |  |
| --- | --- |
|  | 1. Wash your hands. |
| A picture containing text  Description automatically generated | 1. Gather supplies including medication act-o-vial, syringe/needle, and two alcohol pads. |
| A hand holding a small bottle  Description automatically generated with low confidence | 1. Mix the medication by pushing down to release the center stopper. |
| A picture containing indoor, wooden, wood  Description automatically generated | 1. Shake the act-o-vial gently to mix contents. Make sure the solution is clear with no leftover powder. |
| A picture containing gambling house, room  Description automatically generated | 1. Remove the plastic tab from the act-o-vial and clean visible rubber stopper with alcohol pad. |
| A person holding a syringe  Description automatically generated with medium confidence | 1. Remove syringe and needle from packaging. If they are separate, screw needle onto syringe until tight. |
|  | 1. Remove the cap from the needle. Insert the needle directly through the middle of the rubber stopper (see picture in step 5). |
|  | 1. Turn upside down and remove desired dose as directed by medical professional. Push large bubbles back into the vial. The top of the syringe plunger should be level with the dosing line. |
| A baby's foot on a wooden surface  Description automatically generated with low confidence | 1. Select your injection site. Draw an imaginary line from your knee to your hip bone, down the thigh vertically. Draw another imaginary line medially across the thigh horizontally. Those two lines intersect among the outer portion of the middle thigh where you will be injecting. |
|  | 1. Clean site with an alcohol prep pad. |
|  | 1. Use your thumb and index finger to gently pull the injection site taunt. |
|  | 1. Holding syringe like pencil or dart, insert the needle under the skin at 90-degree angle. Hold the syringe steady and inject the medication. |
|  | 1. Pull needle out of skin and discard accordingly. |
| **Go to the emergency room or call 911!** | |