What are the adrenal glands?
Your body has two adrenal glands. Each gland is located above a kidney. The adrenal glands secrete many hormones needed for the body's normal functioning. Two of these hormones are cortisol and aldosterone. Cortisol helps the body use sugar and protein for energy and enables the body to recover from infections and stresses (for example: surgery and illness). Aldosterone maintains the right amount of sodium (salt), potassium, and water in the body.

What is adrenal insufficiency?
People with Adrenal Insufficiency (AI) do not have enough of the hormones cortisol and aldosterone. Without the right levels of these hormones, your body cannot maintain essential life functions. AI may be permanent or temporary.

When AI is permanent, medication must be taken daily for an entire lifetime. Causes of permanent AI include the following:
- Addison’s Disease
- Congenital Adrenal Hyperplasia (CAH) discovered in childhood
- Complete surgical removal of the pituitary gland

Temporary AI is caused by some medications, infections, and/or surgeries. Causes of temporary AI include the following:
- Transsphenoidal surgery for Cushing’s disease that removes a tumor from the pituitary gland
- Removal of a tumor causing the adrenal glands to make too much cortisol
- Medical treatment for Cushing’s syndrome with drugs that lower cortisol levels
- Medical treatment with steroids for prolonged periods of time

What are the signs and symptoms of AI?
When your essential life functions are not being maintained because of a lack of adrenal hormones, you will not feel well. Your symptoms can include:

- Unusual tiredness and weakness
- Dizziness when standing up
- Nausea, vomiting, and diarrhea
- Loss of appetite
- Upset stomach
- Joint aches and pains

Other symptoms you may experience over time include:
- Weight loss
- Darkened skin
- Craving for salt

If any of these symptoms appear and you know that you have a risk for developing AI, call your health care provider immediately.
What medication is used to treat AI?  
To keep your AI under control, you must take medication daily to replace missing hormones. This medication is in pill form and must be taken in the amounts and at the times prescribed by your health care provider. This medication is often referred to as your replacement dose. Many medications can replace cortisol; they are called glucocorticoids. At NIH, hydrocortisone, dexamethasone, or prednisone are usually recommended. If you miss a dose, follow the instructions of your health care provider.

If you are missing the hormone aldosterone, your body cannot maintain the right levels of sodium (salt) and fluids. To replace aldosterone, you will be given a drug called fludrocortisone (Florinef). Adults usually take tablets of fludrocortisone (Florinef). Children with AI who have trouble swallowing pills can take fludrocortisone (Florinef) tablets dissolved in water or crushed. Occasionally, you will be given salt tablets. If you miss a dose of fludrocortisone (Florinef), take it as soon as possible.

What are the side effects of these drugs?  
Replacement doses of hydrocortisone rarely cause side effects. Sometimes, an upset stomach may occur. If this happens, take your medication with meals. If you notice anything else out of the ordinary, call your health care provider. If the dose is too high, patients can gain weight or develop signs of Cushing’s syndrome.

What do I do when I do not feel well?  
There may be times when you do not feel well. When you are sick, be sure to take the right amount of medication at the right time of day. If you feel sick for more than three days, contact your health care provider. There may be times when you will need to take more than your normal replacement dose of hydrocortisone. Normally functioning adrenal glands produce more hydrocortisone when the body is under the physical stress of fever (over 100 degrees Fahrenheit), infection, surgery, trauma with loss of consciousness, vomiting, or diarrhea. It is important to drink plenty of sugar- and salt-containing fluids when you are sick to prevent dehydration or low blood sugar.

If you are sick with a fever (over 100 degrees Fahrenheit), infection, vomiting, or diarrhea, you should call your health care provider right away. Your health care provider may give you written instructions for sick days (“sick day rules”). Generally, this means doubling your usual hydrocortisone dose for 1 to 3 days. It is important to discuss the decision to increase the dose with your health care provider. It is important you increase your hormone dose only for physical stresses. You should not increase it for mental stress (such as a bad day at work, anxiety, or loneliness).

What if I am so ill that I cannot take my medication?  
If you are too ill to take your pills, or you cannot keep them down (for example vomiting), you must take a glucocorticoid medicine by injection. You or someone who lives with you will need to learn how to give you this injection.

The injection will take the place of both hydrocortisone and Florinef pills. If you find it necessary to give yourself an injectable medication, call your health care provider, or go to the nearest hospital emergency room immediately after giving the injection.

You should have the injectable medication with you at all times, make...
sure to check the expiration date periodically. If used, you should replace as soon as possible.

**How much medicine should I take once I feel better?**
As soon as your illness is over and the symptoms are gone (for example, fever, vomiting, and diarrhea), you can usually return to taking your usual amount of medication. You should discuss this with your health care provider.

**How do I give myself an injection?**

Injectable glucocorticoid is given intramuscularly, which means it is injected into a large muscle. When giving yourself an injection, the easiest and best place to do it is in the thigh on the same side as your dominant hand (for example, the right thigh if you are right-handed). Adults should always carry injectable medication with them. If you have a child with AI, you or the child's caregiver must always carry the child's medication. If the child is in school, the school nurse must know about your child's condition and be able to give an injection of glucocorticoid.

How to give an injection of hydrocortisone: (Pictures Courtesy of Kathy Feigenbaum and Jamie Menaker, NIH CCND)

<table>
<thead>
<tr>
<th>Pictures</th>
<th>Directions</th>
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<tbody>
<tr>
<td><img src="image1.png" alt="Image" /></td>
<td>1. Wash your hands</td>
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<tr>
<td><img src="image2.png" alt="Image" /></td>
<td>2. Assemble your equipment</td>
</tr>
<tr>
<td><img src="image3.png" alt="Image" /></td>
<td>3. Mix the medication vial by pushing down on top of the vial to release the cork</td>
</tr>
<tr>
<td><img src="image4.png" alt="Image" /></td>
<td>4. Shake the vial to mix the medication solution well</td>
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<tr>
<td><img src="image5.png" alt="Image" /></td>
<td>5. Use alcohol to clean the rubber stopper on the vial</td>
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<tr>
<td><strong>6.</strong> Take the cap off of the syringe needle. Insert the needle into the vial.</td>
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<tr>
<td><strong>7.</strong> Draw up the medication into the syringe. Adults should use all of the medication in the vial. For a child, use the dose prescribed by the health care provider.</td>
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<tr>
<td><strong>8.</strong> Replace the needle cap.</td>
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<tr>
<td><strong>9.</strong> Select your injection site. To inject yourself safely, become familiar with your body. Undress your thigh, and look at it. Now, draw an imaginary line in the middle of your thigh vertically to divide it in half lengthwise. The outer portion is where you will be injecting. Now, imagine your thigh divided into three equal horizontal rows, from the knee to the hip. The outer portion of the middle third of your thigh is where you will do the injection.</td>
<td></td>
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<tr>
<td><strong>10.</strong> Use alcohol to cleanse the injection site on your skin.</td>
<td></td>
</tr>
<tr>
<td>Step</td>
<td>Instruction</td>
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<tr>
<td>11.</td>
<td>Remove the cap from the needle. Hold the syringe like a dart</td>
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<tr>
<td>12.</td>
<td>Use your thumb and first two fingers to spread your skin while pushing down lightly</td>
</tr>
<tr>
<td>13.</td>
<td>Dart the needle into the thigh injection site, going straight in at a 90-degree angle</td>
</tr>
<tr>
<td>14.</td>
<td>Hold the syringe steady, and inject the medication</td>
</tr>
</tbody>
</table>
15. After injecting the medication, put tissue or gauze near the needle, and pull the needle out quickly

16. Massage the injection site gently

17. Place the syringe and needle in a hard, unbreakable container (such as an empty coffee can with a lid) before disposing of it

18. Call your primary health care provider

### What else do I need to know about AI?

You can control AI by taking an active role in your care. Taking care of yourself involves:

1. Learning about your disease
2. Taking your medication every day
3. Recognizing illness and taking special care of yourself
4. Getting regular medical check-ups

5. Wearing a Medic-Alert bracelet at all times
   - This will be given to you before you leave the Clinical Center. If you are ordering medical alert identification we advise writing on it “adrenal insufficiency, requires cortef”

6. Inform your health care providers of the diagnosis of AI before any surgical procedure to determine whether “stress dosing” is needed.

7. You may want to check in with your local Emergency Medical System to find out about policy/procedures for managing AI in your area.

If you follow the guidelines here and the instructions of your health care team, you will be able to lead a full and productive life. Only you can take care of yourself.

Additional information: CARES Foundation www.caresfoundation.org

**Glossary**

Addison’s disease
An illness caused by the failure of the adrenal glands to secrete enough adrenal hormones

**Adrenal glands**
Two glands, each located above a kidney that secrete cortisol and other essential hormones

**Aldosterone**
A hormone made by the adrenal glands that regulates salt, potassium, and water in the body

**Congenital Adrenal Hyperplasia (CAH)**
A deficiency of adrenal hormones that is present from birth

**Cortisol**
A hormone secreted by the adrenal glands that regulates carbohydrate and protein metabolism

**Cushing’s disease**
A pituitary tumor that makes too much ACTH and causes Cushing syndrome

**Cushing’s syndrome**
An illness caused when too much cortisol is made by the adrenal glands

**Glucocorticoid**
A hormone that is produced by the adrenal cortex that is involved in carbohydrate, protein, and fat metabolism and has anti-inflammatory properties

**Hydrocortisone**
A form of cortisol
This is the drug used to replace cortisol in the body.

**Transsphenoidal surgery**
Highly specialized neurosurgery in which tumors are removed from the pituitary gland.

This information is prepared specifically for persons taking part in clinical research at the National Institutes of Health Clinical Center and may not apply to patients elsewhere. If you have questions about the information presented here, talk to a member of your health care team.

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**Questions about the Clinical Center?**