

Hypopituitarism



IMPORTANT TELEPHONE NUMBERS

Family Doctor

Name: _____

Tel: _____

Pediatrician

Name: _____

Tel: _____

Endocrinologist (Hormone doctor)

Name: _____

Tel: _____ Fax: _____

Endocrine Nurse

Name: _____

Tel: _____ Fax: _____

Pharmacy

Name: _____

Tel: _____ Fax: _____

Emergency Contact Information

Name: _____

Paging: _____

**FOR LOSS OF CONSCIOUSNESS
OR ANY OTHER SEVERE MEDICAL CRISIS,
GIVE INJECTABLE HYDROCORTISONE IMMEDIATELY
AND CALL 911 FOR ASSISTANCE.**

Your Child's Hormone Replacements

1. Cortisol replacement

Daily routine: _____

Illness dosing: _____

EMERGENCIES: Give emergency medicine by injection. Contact your local physician for instructions or go to the hospital.

Emergency dosing: _____

2. Growth hormone replacement

Daily routine: _____

Additional instructions: _____

3. Thyroid hormone replacement

Daily routine: _____

Begin thyroid replacement on _____

4. Sex hormone replacement

Plan: _____

5. Anti-diuretic hormone replacement

Daily routine: _____

6. Other: _____

Note:

Some replacement hormones must be refrigerated.

Check labelling carefully.

Hypopituitarism

“When we first learned that our baby has hypopituitarism, we were overwhelmed.

Now we have settled into a routine – we give him some medicine by mouth and a tiny daily shot – he hardly notices it.

Things are looking up and we are enjoying our baby every day.”

You and your family have just learned that your baby/child has a condition called hypopituitarism. It is very upsetting to find out that your child has a medical condition and will have to take medicines for his or her whole life. This booklet will help you to understand the information that your doctor and nurse have given you. As you learn about the medicines, you will be able to rest assured that your child will have the right amount of hormones in his or her body and will grow and develop like other children. You will be able to relax and enjoy your child for the unique person that s/he is.

This booklet will tell you about hypopituitarism:

- The work that each hormone does.
- Your child’s medicines (which are really hormone replacements).
- How to give the hormones.
- Where you can get further information.



What is Hypopituitarism?

Hypopituitarism is a condition in which the body does not make enough of some or all of the pituitary hormones. The pituitary hormones are command hormones. They direct the activity of other glands or other functions of the body. The pituitary gland itself is under the direction of the hypothalamus.

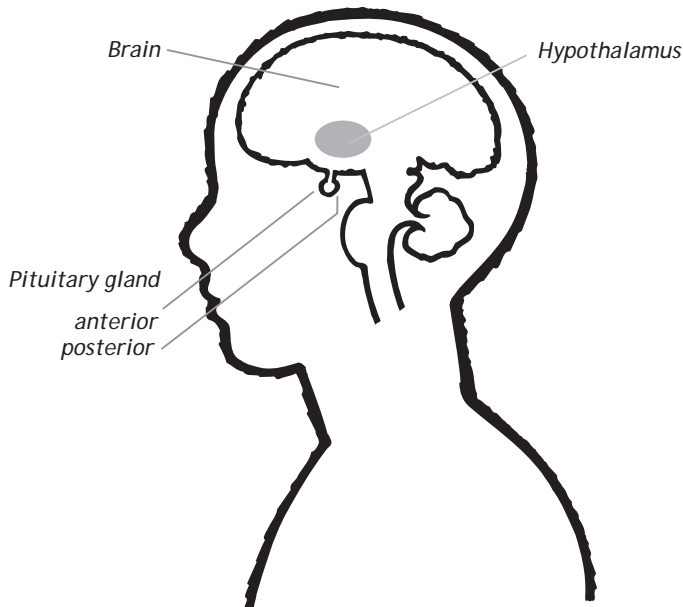
Both the pituitary gland and the hypothalamus are located in the center of the skull, just below the brain. They are often called the command center of the body.

A lack of one or more pituitary hormones is called “hypopituitarism”. A lack of three or more pituitary hormones is sometimes called “panhypopituitarism”.

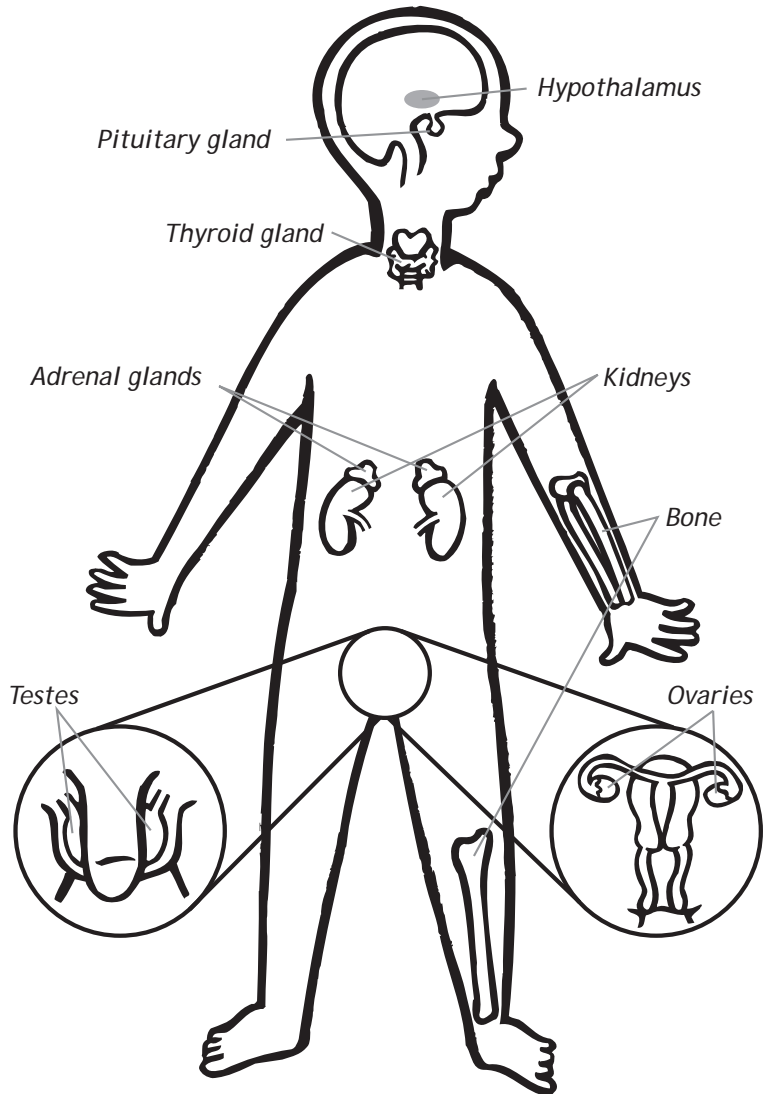
The condition can be present from birth, or your child can lose some or all of the hormones over a period of time.

Hypopituitarism occurs in 1 in 100,000 people. It affects boys and girls equally.

Hypopituitarism is treatable.



Pituitary Hormone System



What are the hormones made in the pituitary gland?

Each one of these hormones will be discussed in more detail in this booklet. This is a summary of all of the hormones. *Ask your doctor or nurse to check off the specific hormones that your child lacks at this time.*

- ❑ **Adrenocorticotrophic hormone (ACTH)** stimulates the adrenal gland to make cortisol. Cortisol circulates in the blood and is responsible for many body functions, such as keeping the blood pressure stable and keeping the blood sugar in the correct range. Cortisol is necessary in the body's response to stress.
- ❑ **Growth hormone (GH)** is made in the pituitary gland. It circulates in the blood and influences the production of a chemical called insulin-like growth factor-1 (IGF-1) in the liver and other cells in the body. GH and IGF-1 stimulate the growth plates in bones and growth in other tissues.
- ❑ **Thyroid stimulating hormone (TSH)** stimulates the thyroid gland to produce the thyroid hormones. Thyroid hormones circulate in the blood and influence the work of every cell.
- ❑ **Luteinizing hormone (LH)** and **follicle-stimulating hormone (FSH)** stimulate the ovaries and testes (testicles) to produce estrogen, progesterone and testosterone - the sex hormones. The sex hormones cause the changes of puberty - breast and body hair development and menstruation in girls, and penis and testicular enlargement and body hair development in boys. LH and FSH also cause the ova (eggs) or sperm to mature.
- ❑ **Antidiuretic hormone (ADH)** is made in the hypothalamus and in the posterior (back) pituitary gland, and causes the kidneys to keep the right amount of water in the body.

Your child may lack one or two hormones right now. It is likely that s/he will continue to be deficient in those hormones.

However, it is also possible to become deficient in some of the other pituitary hormones in the future.

What causes hypopituitarism?

There is no known way to cause incomplete pituitary development.

It didn't happen as a result of anything you did.

Hypopituitarism may be present from birth if the hypothalamic or pituitary area did not develop properly in the womb. Other problems of fetal development (development before birth) may be present as well, such as optic nerve hypoplasia (incomplete development of the nerves to the eyes) or cleft lip and palate (incomplete closure of the roof of the mouth and the upper lip).

Hypopituitarism may happen later in life as a result of disease such as histiocytosis; a head injury; infection, tumour, or surgery in the pituitary area; or radiation to the head.

How does the doctor know my child has this condition?

Your newborn may be checked for this condition if his/her body shows the result of lack of hormones. For example,

- Low blood sugar in a newborn may mean that the baby is not producing enough growth hormone or cortisol.
- A small penis in a boy baby may mean that the baby is not producing enough growth hormone or testosterone.
- Prolonged jaundice in an infant may also indicate hypopituitarism.

Older children may show signs of

- poor growth,
- low blood sugar,
- lack of timely pubertal development,
- signs of lack of cortisol – tiredness, headaches, nausea, slow recovery from illness.

Blood tests are done to measure the amount of hormones in the child's body. Sometimes the tests show that the body is making some of the hormone, but not enough to do the job.



Children who have had tumours, surgery or radiation in the pituitary area of the brain will be watched for hypopituitarism.

Imaging tests are done such as computerized tomography (CT) scans or magnetic resonance imaging (MRI) to look at the pituitary and hypothalamic area of the brain. These tests may help to explain why your child has a lack of hormones.

How is the condition treated?

Replacement hormones are needed throughout your child's life – just like the ones nature makes.

The hormones that are missing can be replaced in a way that is as much like nature as possible. You can usually expect normal growth and development in your child. However, sometimes the condition which has caused the hypopituitarism has other effects on your child's body. For example, optic nerve hypoplasia sometimes causes problems with vision and intellectual development. Radiation for a brain tumour may also cause other problems.

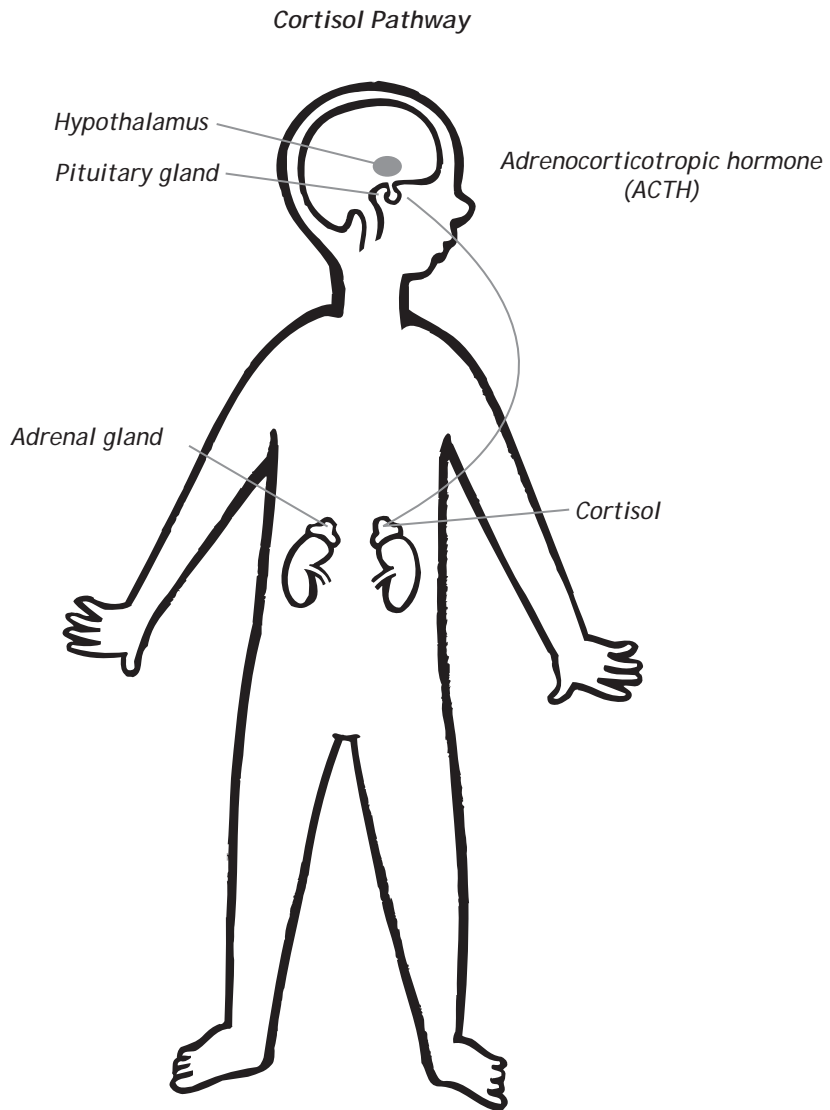
In the next sections of this booklet, each hormone will be described – how it works, how it is replaced, and how the health care professional makes sure the dose is correct.

Check back on the list of pituitary hormones to know which sections apply to your child.

Cortisol Replacement



Cortisol Replacement



Cortisol Replacement.

Cortisol is a life-sustaining hormone:

- It keeps the blood pressure in the right range.
- It keeps the blood sugar in the right range.
- It makes sure the cells have enough glucose for their activity by breaking down protein and fat when needed.
- It helps the body deal with stress.

Your child's adrenal glands (where cortisol should be made) are perfectly healthy, but they are not receiving a command from the pituitary gland to make the hormone cortisol, so it must be replaced.



Cortisol is like gas in a car. It enables the engine to run.

How does the doctor know that our child lacks cortisol?

When the doctor suspects that your child is deficient in cortisol, a blood test will be done. An intravenous is started, medicine similar to the pituitary-stimulating hormone ACTH is given, and several small samples of blood are taken. The results of this test will show whether your child makes enough cortisol for daily needs and also for special situations such as illness.



How is cortisol replaced?

Cortisol is replaced by a medicine from the glucocorticoid family. The pill or syrup is given by mouth. One of the following will be recommended:

- hydrocortisone, also called Cortef®
- prednisolone, also called Pediapred®
- prednisone
- dexamethasone, also called Decadron®

Each is slightly different in their power – for example prednisone is 4 times more powerful than hydrocortisone. Each is different in how long it lasts in the body. The doctor chooses the one that s/he feels is best for your child.



Note: Always check the product that your pharmacist has given you, because many of the names sound similar.

The most common glucocorticoid for children is hydrocortisone. It is recommended for children because it is most like nature, and it provides the best opportunity for the child to grow well. It is usually taken 3 times each day because the body uses it up fairly quickly. It may be irritating to the stomach so many children take it with food, formula or milk.

Cortisol dosage does not need to be increased for emotional stress

The doctor determines the dosage based on your child's height and weight, and your reports of how your child is feeling. The doctor will also watch your child's growth carefully:

- Too much weight gain at the same time as poor growth may mean that the dosage is too high.
- Lack of usual energy and unusual tiredness may mean the dose is too low.

Concerns about your child's growth and energy level can be discussed at your regular appointments.

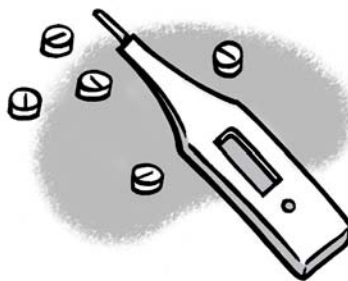
How is cortisol replaced during illness or injury?

Management Instructions in the Appendix of this booklet give additional details and provide space for specific dosage directions for your child.

When we are sick or injured, our pituitary gland tells the adrenal gland to make lots more cortisol. The extra cortisol helps the body with all its activities in coping with illness. Your child's body is not able to make more as needed, so you will have to increase the dose.

Give the increased dose when the child is sick with

- infection
- fever over 38.5°C or 101°F
- moderate trauma such as stitches



An infant with a fever should be seen by a doctor

How is cortisol replaced during an emergency?

In an emergency, hydrocortisone must be given by intramuscular injection by the parents. After the injection, the child should go to hospital to be seen by a doctor. The injection will give the child enough hydrocortisone for a short time (approximately 6 hours), but the illness itself may need treatment.



Emergency situations include:

- vomiting
- diarrhea
- fracture of a major bone
- major seizure
- loss of consciousness

Hydrocortisone must be given by injection (intramuscular or intravenous) every six hours, until your child is able to take the appropriate dose by mouth.

In case of general anesthetic and surgery, ask your endocrinologist (hormone doctor) and your anesthetist to plan the amount of injectable hydrocortisone to be given before the surgery and during recovery.

Instructions in the Appendix of this booklet will describe how to give an intramuscular injection

Giving hydrocortisone when it is needed will prevent your child from having a lack of cortisol in his/her body. When the body lacks cortisol, it is called an **adrenal crisis**.

If there is not enough cortisol in your child's body, s/he will go into an adrenal crisis – weakness, stomach pain, nausea, vomiting, seizures, and loss of consciousness.

**An adrenal crisis is a medical emergency.
It is life-threatening.**

*Remember:
extra hydrocortisone
given for a short time
for illness will not cause
any bad effects, but lack
of cortisol will cause
serious adrenal crisis.*

The goal of treatment is to always have enough cortisol in the body – just like it is a goal to always have enough gas in the tank. In that way, adrenal crisis (running out of gas) is prevented.



*Cortisol is like gas in a car.
If there is not enough gas, the car won't run.*

Are there any side effects from cortisol replacement?

Synthetic hydrocortisone is the same as cortisol which is made in the body, so side effects are not expected. The amount that is prescribed for your child is the amount needed for his or her age and size. Occasionally the dose is slightly too much and the extra amount will cause decreased growth, increased appetite and weight gain. Your health care professional will recognize this at your regular appointments and adjust the dose.

When should I call the health care professional?

Sometimes it is difficult to decide whether your baby needs an increased dosage. Call your health care professional to discuss the situation, but if you are unable to contact anyone, give the illness dosage just in case.

Remember the emergency situations. If your child is in any of these situations, give the injection then take your child to the hospital.



Emergency situations include:

- vomiting
- diarrhea
- fracture of a major bone
- major seizure
- loss of consciousness

Note: It is necessary for children with cortisol deficiency to wear a medical alert bracelet. This will tell health care providers about your child's condition in the event of an emergency.

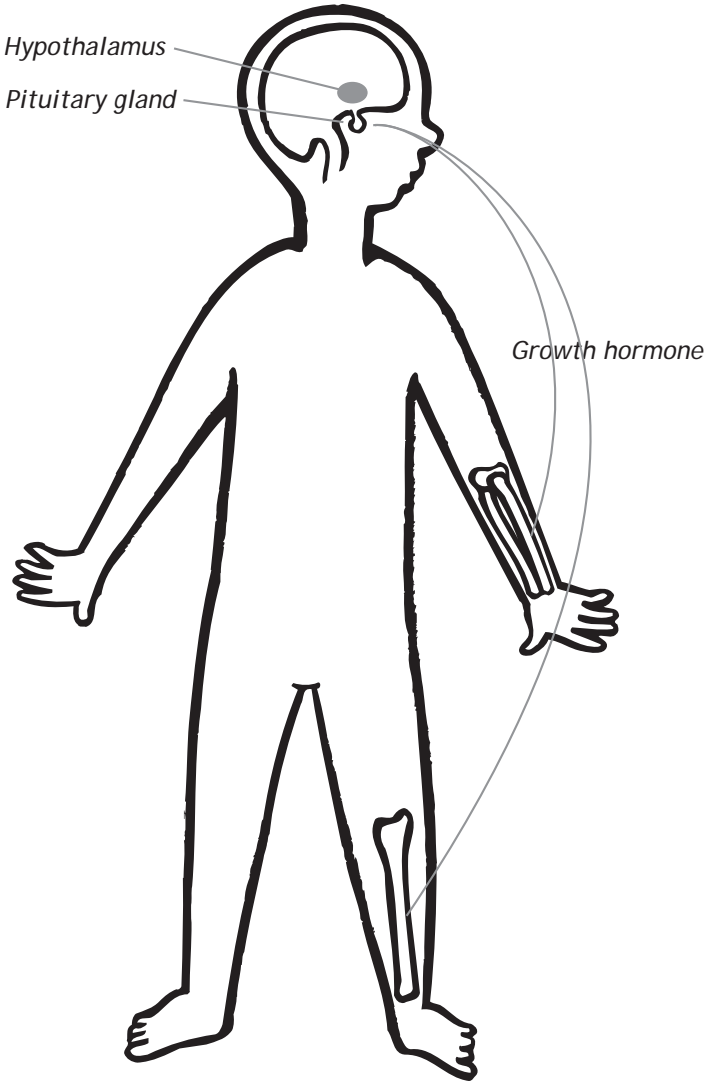


Growth Hormone Replacement



Growth Hormone Replacement.

Growth Hormone Pathway



Growth Hormone Replacement

Growth hormone is made and stored in the pituitary gland. It has several important jobs in the body:

- It works with another hormone, insulin-like growth factor-1, to promote the growth of bones and other tissues.
- It causes glucose to be made as needed, so that your child's blood sugar stays in the normal range even when he/she has not eaten for a while.

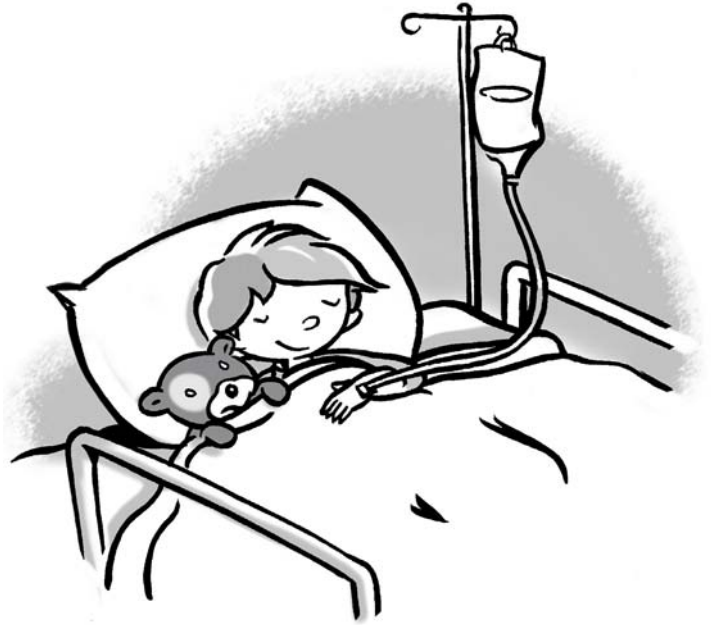
How does the doctor know that my baby/child lacks growth hormone?



Something will have happened to make the doctor think that your child does not have enough growth hormone:

- low blood sugar
- lack of other pituitary hormones
- poor growth in an older child.

A blood test called a “growth hormone stimulation test” will be done. This is a long test (about 4 hours). An intravenous will be started. Several different medicines will be given to your child to make the pituitary gland release stored growth hormone into the blood. Then the nurse will use the same intravenous to take out tiny blood samples for analysis. The blood test results will show if your child is making enough growth hormone.



How is growth hormone replaced?

Growth hormone is replaced using synthetic growth hormone given by daily injections into the fatty layer just below the skin. Although children do not like injections - and parents do not like to give injections - this is the only way to get the growth hormone into the body. It would be destroyed in the stomach if it were given by mouth. Parents learn how to give the injection themselves. Your child may want to learn to do it when s/he is older. There are several injection devices available to make it more convenient to give injections. You can ask your nurse for more information about injection devices.



The amount of growth hormone is usually determined by your child's weight. As your child gets older his weight increases and the dosage will increase.

Growth hormone will be replaced during the growing years. When growth is finished (approximately age 14-17), you will discuss whether retesting or ongoing treatment is needed with your health care professional.

If your baby has had low blood sugar, it is very important to give the injection daily, preferably in the evening.

If your child is older and does not have low blood sugar problems, the occasional dose may be missed for sleepovers or other events.

Talk with your health care professional about this.

Instructions in the Appendix of this booklet will describe how to give a subcutaneous injection

Are there any side effects from growth hormone replacement?

Synthetic growth hormone is the same as the natural growth hormone the body should be making, so side effects are not expected. The doctor will watch for possible rare side effects of growth hormone:

Most growth hormone products must be refrigerated.

- increased pressure in the head (frequent headaches and vomiting)
- increased sugar in the blood
- joint problems

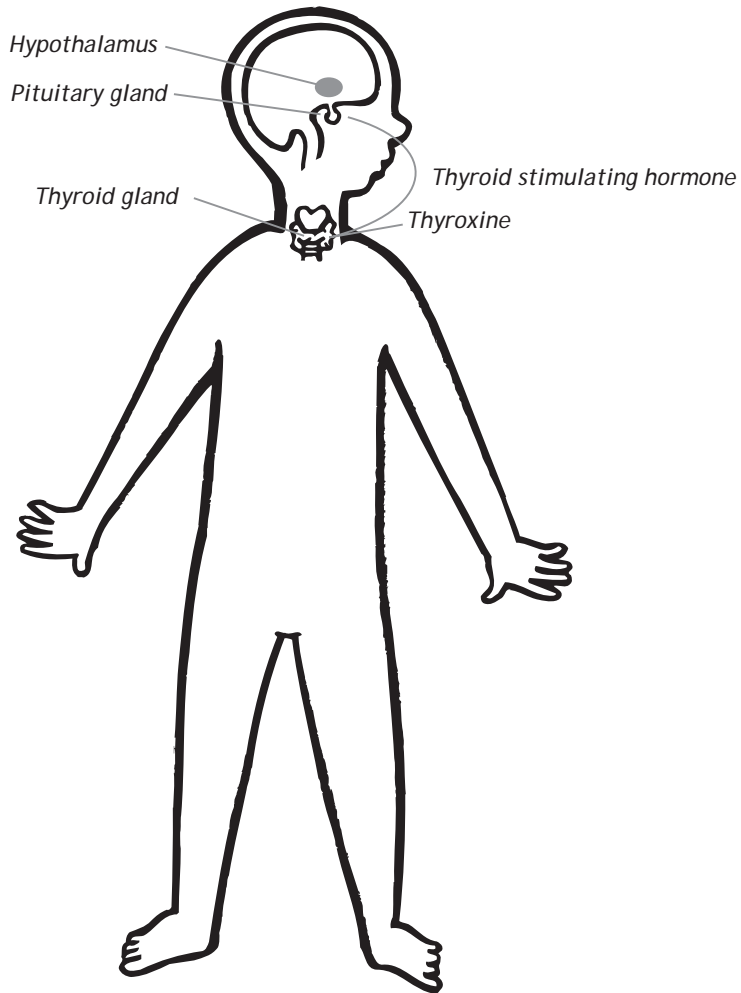
Parents of children who have had tumours or radiation are very concerned about the tumour coming back. The information available at this time shows that growth hormone does not increase the chances of the tumour coming back. It also does not increase the chances of new tumours developing. Your doctor will be able to give you more information about your child's situation.

Thyroid Hormone Replacement



Thyroid Hormone Replacement

Thyroid Hormone Pathway



Thyroid Hormone Replacement

The thyroid hormones are important for growth and development. They are necessary for normal brain development. The thyroid gland in your child's body is perfectly healthy, but it is not getting the command from the pituitary gland, so the thyroid hormones are not being made.

How does the doctor know that our child lacks thyroid hormones?

When the doctor is concerned that your child may have hypopituitarism, a simple blood test is done to measure the thyroid hormones.



My Mom puts EMLA® cream on my arm an hour before the blood test. It numbs a little spot on my arm so the poke doesn't hurt.

How is thyroid hormone replaced?

Thyroid hormones are replaced by synthetic thyroxine, a small sweet-tasting tablet taken by mouth. At first the dose is determined by the age and weight of your child, and then adjusted according to the levels of thyroid hormone in the blood. This means that your child will need a small amount of blood taken – every three months at first, and later on, every 6-12 months. This is a replacement hormone, so the dosage will increase as your child grows.



Note: Thyroid hormones will increase the metabolic rate of all the cells' activities. Cortisol is necessary for metabolism, so if both hormones are missing, cortisol must be replaced for several days **before** beginning thyroxine replacement.

Are there any side effects from thyroid hormone replacement?

Synthetic thyroid hormones are the same as thyroid hormone made in the body, so side effects are not expected.

- If the dosage is too high, your child will become “hyperthyroid”, resulting in a fast pulse, sweaty skin, inability to sleep, weight loss and poor concentration at school.
- If the dosage is too low, or your child misses frequent doses, your child will become “hypothyroid” which results in tiredness, sluggishness, weight gain, constipation, and poor growth.

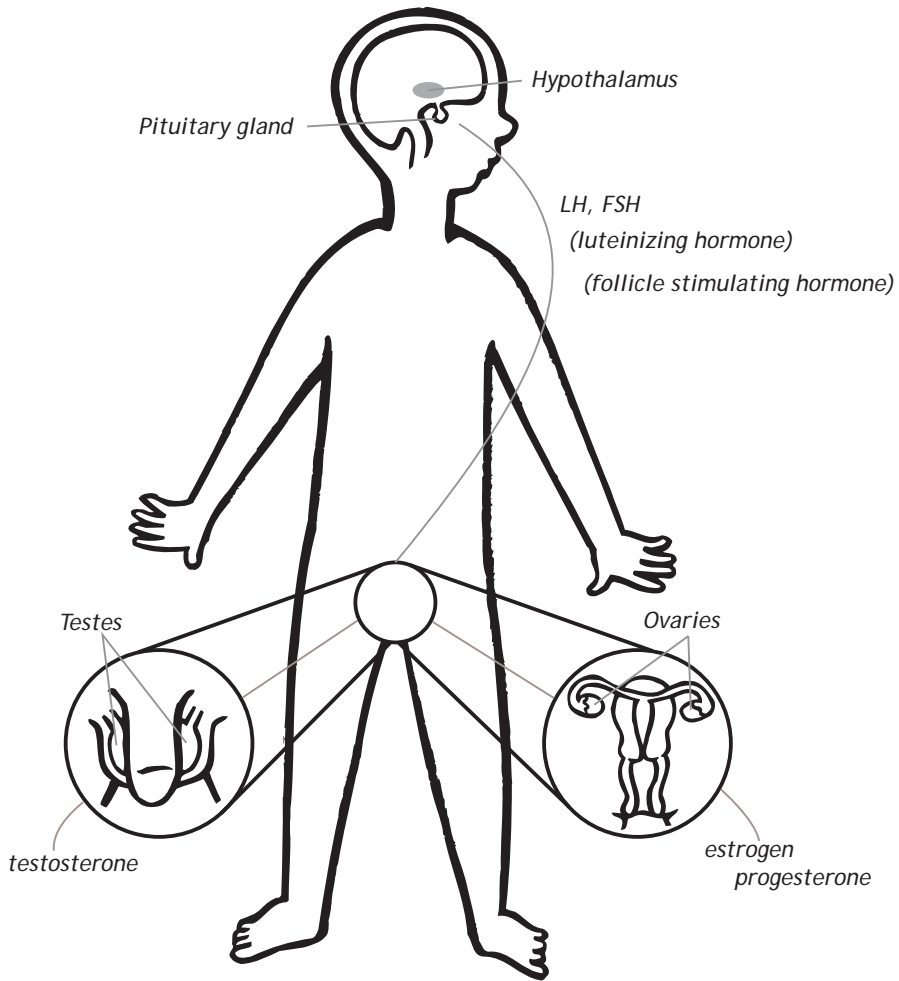
This is unlikely to happen because your child is taking thyroid hormone replacement, and regular blood tests will be done to adjust the dosage as needed, keeping the thyroid hormone in the right range.

Sex Hormone Replacement



Sex Hormone Replacement.

Sex Hormone Pathway



Sex Hormone Replacement

“Sex hormones” is the term used for the hormones made by the ovaries or testes. When the pituitary does not make adequate amounts of luteinizing hormone (LH) and follicle-stimulating hormone (FSH), then the ovaries and testes do not do their job in making the appropriate hormones. This means that there will not be enough estrogen and progesterone in girls, and testosterone in boys. The ovaries or testes in your child’s body are healthy, but they do not get the “command” from the pituitary, so the sex hormones are not produced at the normal time of puberty.

Men’s and women’s bodies make all of the sex hormones, but in different amounts. Men make mostly testosterone, and women make mostly estrogen, but each makes some of the other hormone.

How does the doctor know that my child lacks sex hormones?

When your child is young, the doctor may not be able to tell if he or she will produce enough sex hormones to go through puberty in the future. This is a situation of “watchful waiting”. By the age of 12-13, the effects of the sex hormones should have begun to produce changes in the blood levels of the hormones, and changes in the child’s body. If this does not happen, the doctor will discuss replacement hormones with you and your child.

How are the sex hormones replaced?

The hormones are replaced in a manner that is as much like nature as possible. When your child is at the age of puberty, about 12 or 13, the appropriate hormone is replaced at a very low dosage – just like nature. The dosage is gradually increased over several years, until the normal adult dosage is being given.

Testosterone taken orally (by mouth) is not recommended because of the potential for liver damage from the oral medicine.



Hormone replacement for boys:

Boys born with a small penis are often given a small amount of testosterone for a couple of months during their infancy to cause some growth of the penis, just as nature does during infancy.

Beginning at about age 12-13, boys take small doses of

testosterone every 3-4 weeks given by injection into the muscle. The testosterone may also be given in the form of a gel or patch that is applied to the skin. The gel or patch is applied daily, and is much more costly than injections. Your family will decide on the best method of taking testosterone after discussing the advantages of each method with your health care professional.

The dosage is gradually increased over a period of several years, and eventually the testosterone injection is given every 2 weeks.

The testosterone that your teen receives will have the same effects as testosterone made in the body:

- increased growth
- increased size of penis
- development of body hair
- lowered voice
- acne
- muscle development
- sexual interest

Since the testes are not making the testosterone, they will remain small.

Your child will take testosterone for the rest of his life.

Many parents learn to give the injection, and in time, young adults learn to give the injection themselves.

Instructions in the Appendix of this booklet will describe how to give an intramuscular injection

Hormone replacement for girls:

Beginning at about age 12 or 13, girls take small doses of a form of oral (by mouth) estrogen every day. The dosage is gradually increased over a period of several years. As the dosage approaches the adult amount, the doctor will recommend adding the second female hormone, progesterone, for part of each month. Then for about a week each month, no hormones will be taken. This schedule mimics nature, and a menstrual period will begin about 3 days after the hormones are stopped each month.

The female hormones will have the same effects as those normally made in a girl's body:

- increased growth
- breast development
- maturation of the uterus
- menstrual periods
- mood changes
- sexual interest

The amount of body hair that develops may be limited.

The hormones will be taken until the age of normal menopause, and possibly longer.



What about fertility - will my child be able to grow up and be a mother/father?

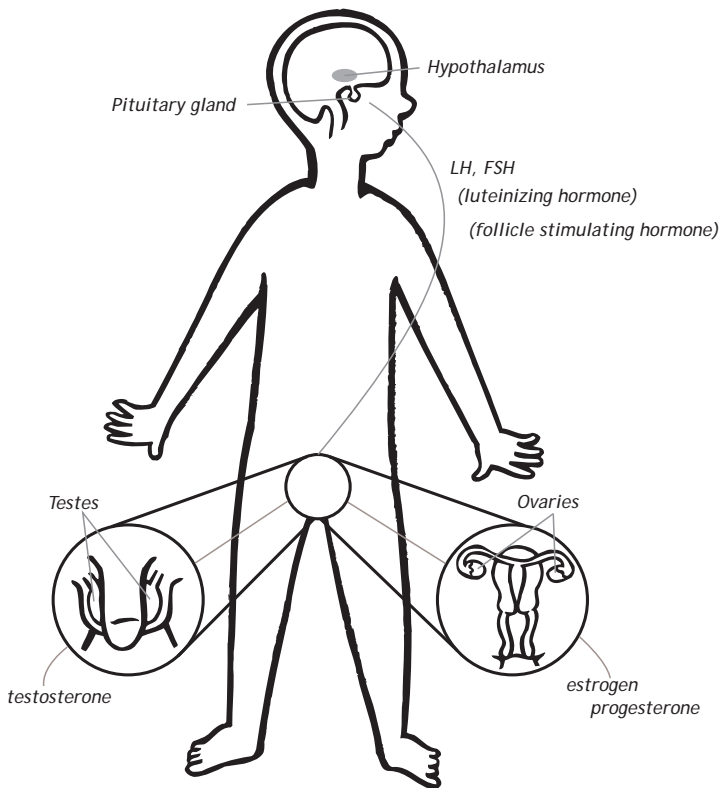
If your child is lacking in sex hormones, they will be replaced starting in the teen years, and your child will have all the changes of puberty. The hormone replacement does not cause eggs or sperm to mature. That means that your child will not be fertile under normal circumstances. However, over the past decade, huge developments have occurred in “reproductive technology”. It is now possible to stimulate the young person’s own testicles or ovaries in such a way that the sperm or egg is matured. This technique has already proven successful in some situations. You can anticipate that reproductive technologies will become even more successful by the time your child is an adult.



Early Puberty (Central Precocious Puberty)

Some children with hypopituitarism may also experience precocious puberty, which means pubertal changes before age 8 in girls, or age 9 in boys. The same disturbance to the pituitary area which has caused hypopituitarism can also trigger early puberty. Parents and/or physicians will know this is happening when they see breast development in girls or pubic hair beginning for either girl, or boy, at an age that is earlier than expected.

Sex Hormone Pathway

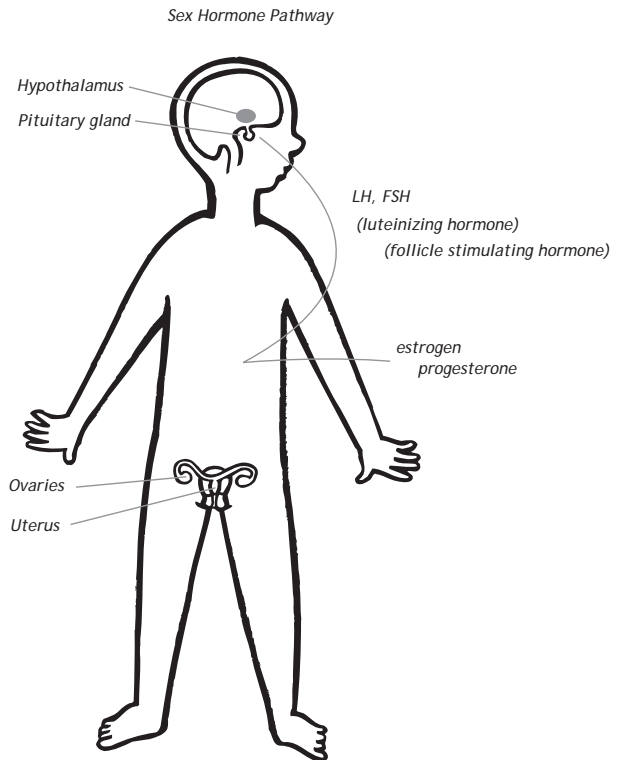


Although puberty may come very early for your child, s/he may still develop deficiency of sex hormones several years later.

Treatment is often recommended, because puberty that comes too early can affect growth and can be difficult for your child socially. A number of medicines can be used. One is Lupron Depot®, a medicine similar to the hormone made in the hypothalamus which is stimulating the development. The medicine is long-acting and works by stopping the body from responding to its own natural hypothalamic hormone. This medicine is given by injection every 1-3 months. Some girls have a light menstrual period a few days after the first injection. You can help your daughter to be prepared for this possibility. Lupron Depot® will be given by injection until the time that puberty is normally expected to occur, about age 12. Pubertal development will begin again when the medicine is stopped, but it is possible that sex hormone deficiency will occur later.

Some families learn to give the injection themselves.

Instructions in the appendix of this booklet will describe how to give an intramuscular injection.

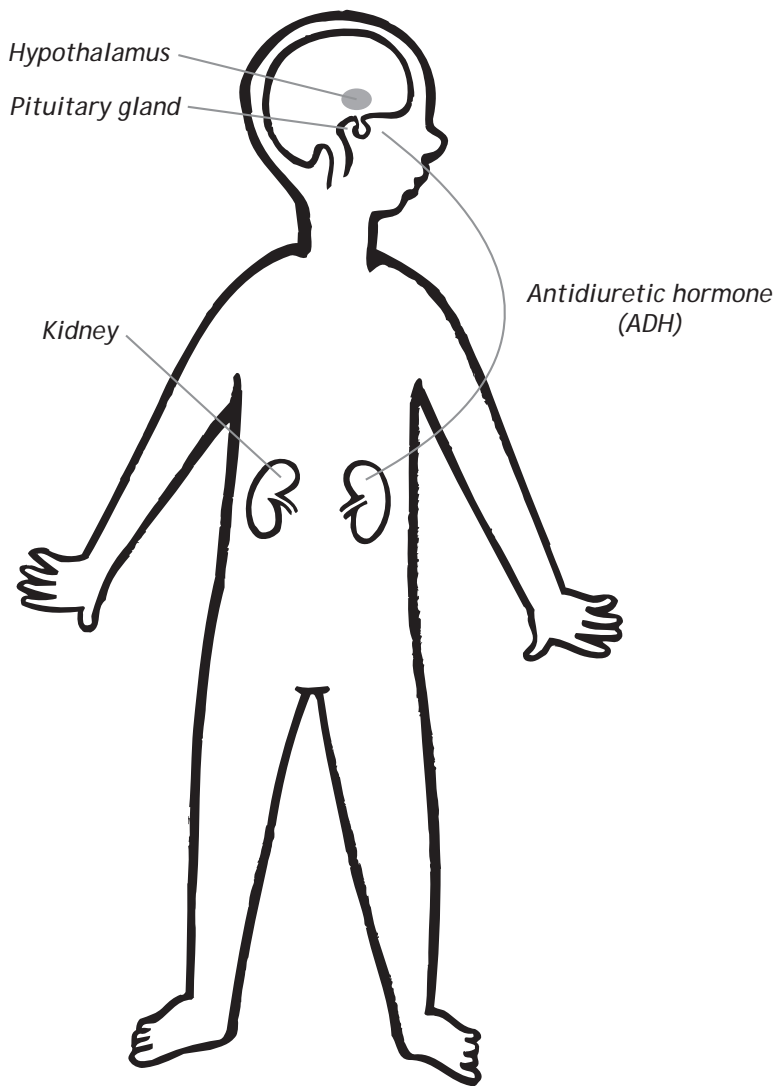


Antidiuretic Hormone Replacement



Antidiuretic Hormone Replacement

Antidiuretic Hormone Pathway



Antidiuretic Hormone Replacement

Antidiuretic hormone (ADH) is made in the hypothalamus and stored in the back part of the pituitary gland. It is released as needed and causes the kidneys to keep the right amount of water in the body. Your child's kidneys are healthy, but they are not getting the command from the pituitary gland. This condition is called central diabetes insipidus (DI).

Diabetes insipidus is not sugar diabetes.

Your child does not have diabetes mellitus (sugar diabetes).

How does the doctor know that my child does not make enough ADH?

Children who do not have enough ADH lose too much fluid in their urine. As a result they are very thirsty, drinking very large amounts and urinating more than normal. When this is happening, tests are done to measure the concentration of blood and urine. This will tell the doctor if your child is making enough ADH.



Children can become very thirsty during this test. This is very uncomfortable for the child but there is no other way to find out if your child has DI.



Sometimes a water-deprivation test is requested. For this test, your child will go without food or fluids, including water, for a period of time. An intravenous will be started and samples of blood and urine will be taken during the test. Measurements will be done on the samples which will tell whether the child's body is producing ADH.

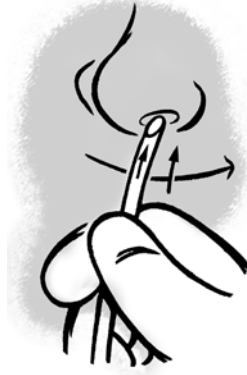
How is antidiuretic hormone replaced?

Antidiuretic hormone is replaced with a synthetic hormone, desmopressin acetate, also called DDAVP®. DDAVP® is given into the nostril, using a tube or spray, or it may also be given by tablet taken by mouth.

You and your doctor will discuss a plan for replacement. At first, the dose is small, and you will watch for the effect.

It is expected to make the child's body keep fluid in, so the child will not be urinating so frequently.

When the DDAVP® has worn off, frequent urination begins, and it is time for another dose. The period of time when the DDAVP® has worn off is called "breakthrough". Based on your reports of breakthrough, the doctor will increase the dose slowly until the DDAVP® lasts for about 12 hours.



Talk with your health care professional if you feel the amount of DDAVP® needs to be adjusted. There are many factors to consider.

Regular blood tests will be needed to be sure the right amount of fluid is being held in your child's body.



Some DDAVP® preparations must be refrigerated. Check the labelling carefully.

Special notes to parents of infants:

Your doctor needs a lot of accurate information to decide on the right dose of DDAVP® for a baby who can't tell you about thirst.

The doctor may ask you to measure and record:

- The amount of fluid that your baby takes in: milk, water, juice and food. The dietitian can show you how to measure the amount of fluid in baby foods.
- The amount of fluid that your baby puts out. The nurse will show you how to measure by weighing diapers on a kitchen scale.
- Urine specific gravity (SG) using a test strip. (A cotton ball is placed in the diaper in the area that becomes wet with urine. When wet, the cotton ball can be pressed on to the test strip and the SG is determined using the colour chart.)
- Daily weight of your baby measured at the same time of day, preferably wearing no clothing.

Your health care professionals will teach you how to use the above information to decide if breakthrough has occurred and another dose of DDAVP® is needed.

Your doctor may also ask you to have regular blood tests to be sure the dose of DDAVP® is just right for your baby.



Are there any side effects of DDAVP®

Most people do not have any side effects from DDAVP®, although from time to time your child may have the effect of too much or too little DDAVP®.

- The effect of too much DDAVP® is that too much fluid is kept in the body – this can cause headaches and stomach ache. If this happens, your child is not given drinks until breakthrough occurs. Then the dosage is begun again at a lower amount.
- The effect of too little DDAVP® is that “breakthrough” occurs very soon after the dose.

This may happen if the:

- dose is too small, or the
 - medicine is too old, or
 - your child has a cold and is not absorbing enough medicine through his/her nostrils. Your health care professional will likely tell you to give another small dose in this situation.
- Some children find nasal DDAVP® irritating to the nostril causing redness or soreness.

*Children with DI
drink to satisfy thirst.*

*Teachers should
give easy access
to bathrooms and
drinking water.*

For a quick summary of all the medicines your child needs, look back to the first pages of this booklet.



Summary

As you can see, there is a lot to learn about the hormones that your child needs. The information may be overwhelming at first, but you will soon settle in to a routine of giving the hormones and enjoying normal life with your child.

The following information answers some of the questions that parents commonly ask. As well, you will find instruction sheets about how to give an injections, and instructions about illness management.

As always, discuss your concerns and questions with your health care team.



Questions and Answers



Questions from Parents

Q: Will this condition go away?

A. Almost always the condition is life-long.

Q: How do you give pills to a baby?

A. Crush with a pill crusher. Dissolve it in ½ teaspoon of breast milk, formula, or sterile water. Pour the mixture into a small spoon, or draw it up into an oral syringe or dropper. Then give this to the baby.

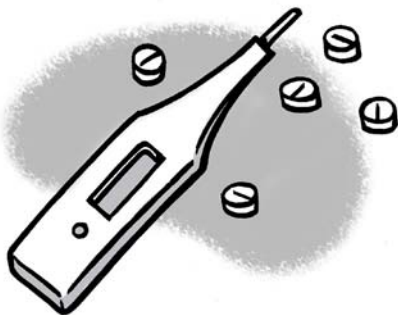
Do not put the pill into a bottle.

The baby may not take the whole dose. Different pills can be given at the same time.



Q: How do I know if my baby is sick and needs extra hydrocortisone?

A. A change in appearance and a change in behaviour are signals to tell you to watch the baby more carefully and check his or her temperature with a thermometer. Your baby may have pinker cheeks, watery eyes, runny nose, watery stools. Your baby may be fussy and refusing to feed as usual. If your baby shows an increase in his temperature, give extra hydrocortisone according to your instructions.



Sometimes the extra hydrocortisone is **not** needed.

For example:

- Your baby has a runny nose and refuses solid food, but is taking fluids, and the temperature is not above normal
- Your baby has had a cold with a fever. You increased the hydrocortisone during the fever. Now your baby's temperature is normal (no fever).
- Your baby has a frequent cough, but he/she is feeding well, sleeping and playing as usual.

In these situations, extra cortisol is not needed, because the baby is not physically stressed by the runny nose or cough.

A baby under 4 months old should not get a fever. If s/he does develop a fever, s/he must be seen by a doctor soon to determine the cause and the appropriate treatment. Also of course s/he needs extra hydrocortisone.

Q: Does it matter when my child takes the hormones?

A. Each hormone has a different answer because each hormone lasts a different length of time.

- Cortisol replacement (hydrocortisone, Cortef®, Pediapred®, prednisone) should be spaced out around the clock – every 8 hours, or every 12 hours as directed. When the child is older, it may be given early morning, late evening before bedtime, and about half-way in between. Illness replacement starts as soon as the illness is recognized, even if it is not a regular dose time.
- Growth hormone is generally given before bed. This mimics nature – growth hormone is usually released during sleep.

- Thyroid replacement can be given any time of day, but a regular time is preferable.
- Sex hormone replacement for girls can be given any time of day. Injections for boys are about every 4 weeks, although a few days more or less would be acceptable.
- DDAVP® is given as needed. The times may be different for each child. It is generally given when “breakthrough” occurs – when the last dose is worn off.

Q: What if I forget to give the hormones for a whole day?

- A. Give a double dose of cortisol replacement and thyroid replacement as soon as you realize they have been missed.

Give the normal dose of other hormones.

Q: Should my child be immunized?

- A. Immunize your child with the routine childhood immunizations on the regular schedule. Immunizations against other communicable diseases such as chicken pox are also recommended. Give your child acetaminophen one hour prior to the immunization. This may prevent a fever. Many doctors also recommend one extra dose of cortisol replacement one hour prior to the immunization. If your child gets a fever, treat him/her according to your “illness management” instructions.

The whole family should have the flu shot every fall.



Q: My baby had low blood sugar. Do I need to worry about this?

A. Your doctor will tell you whether you need to check the blood glucose using a monitor. Usually when babies receive the hormones they need, there is no longer a problem with blood sugar levels.

Q: How much do all these hormones cost?

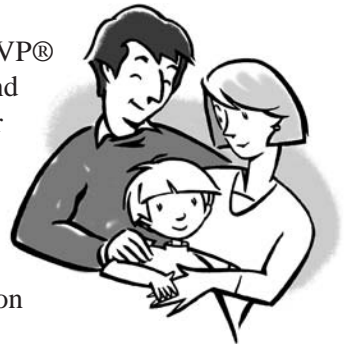
A. The cortisol replacement, thyroid replacement and sex hormone replacements are quite reasonable. DDAVP® costs about \$50 each month, depending on how much your child needs. Growth hormone is very expensive \$10,000 to \$40,000 per year. In Canada, most of the cost of growth hormone is paid by the provincial drug plans. Talk with your health care professional if expenses are causing you concern.

Q: How will my child manage this condition at school?

A: Children with hypopituitarism go to school like all other children. Your child's teacher and the school staff need to understand how to manage your child when he/she gets ill or injured. Generally the school does not need to learn to give emergency injections. They will call you or the Emergency Medical Service to deal with vomiting or a significant injury.



If your child also takes DDAVP® the school needs to understand that s/he needs to drink water when thirsty and use the bathroom as needed



A medical alert bracelet will give essential information in an emergency.

Appendix

MANAGEMENT OF HYDROCORTISONE REPLACEMENT

Hydrocortisone is needed by the body to maintain normal blood pressure and blood sugar. It is therefore necessary for life.

Normal Daily Replacement:

Your child has been prescribed: _____
and should take this as follows:

Morning _____ Afternoon _____ Evening: _____

If a dose is forgotten, take the dose as soon as possible. If the whole day's replacement is forgotten, take a double dose as soon as possible and resume normal schedule.

Stress Dose (Illness Dose)

Your child's body needs a large amount of hydrocortisone during times of physical stress such as:

- Fever more than 101°F or 38.5°C
- Illness such as nausea, throat or ear infection
- Moderate trauma such as an injury requiring stitches or a broken bone

As soon as the illness or injury is recognized, the normal dose should be increased to: _____

If this stress dose is given for more than a week, discuss a plan with the doctor for tapering the dose back to normal.

Special instructions for infant: When a baby is sick, extra precautions should be taken:

- Check the child for fever and treat it as instructed by your doctor
- Be sure the baby gets enough fluid. Give Pedialyte® with 1 teaspoon added sugar, or formula.
- A baby who is not responding normally should be seen by a doctor.

EMERGENCY INJECTION FOR

- Vomiting
- Diarrhea
- Fracture of a major bone
- Loss of consciousness
- Major seizure
- General anesthetic

Hydrocortisone
_____ mg, or _____ ml (cc)
by intramuscular injection.

This injection will act quickly and will last approximately 6 hours. After 6 hours, the child needs more hydrocortisone and if unable to take pills, the injection must be repeated.

Additional instructions: _____

If your child is ill and requires injectable hydrocortisone, s/he will likely need further medical attention. Take your child to the local hospital without delay for further assessment and treatment.



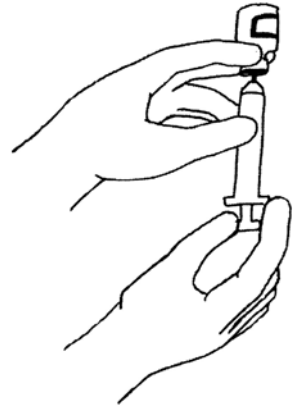
GIVING A SUBCUTANEOUS INJECTION

Medication: _____

Supplies: _____

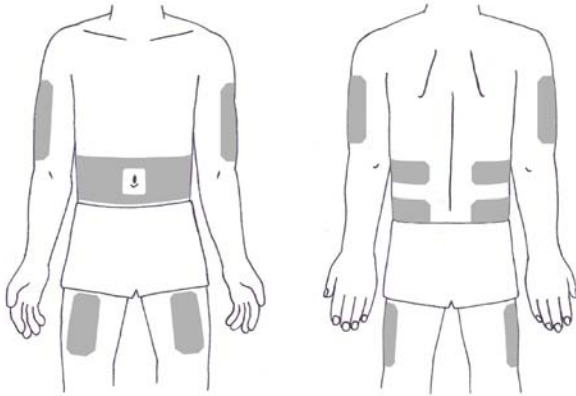
Drawing Up the Medication:

1. Wash your hands thoroughly
2. Check that the medication is correct and not past the expiry date. If the medication will be used for more than one dose, record the date of the first dose on the label. Be aware of the maximum length of time that the vial can be used.
3. Clean the top of the bottle with an alcohol swab.
4. Remove the needle protector. Do not let the sterile needle touch anything.
5. Pull back the plunger, drawing up the amount of air equal to the amount of medicine to be given.
6. Insert the needle into the rubber stopper on top of the bottle. Push the air into the bottle.
7. Turn the bottle and syringe upside down.
8. Pull down on the plunger, drawing up the correct amount of medicine. The end of the needle must be in the medicine to draw up medicine.
9. While the needle is in the medicine, look for large air bubbles. Remove them by pushing them back into the bottle. Recheck that you have the correct amount of medicine.
10. Carefully re-cap the needle to prevent contamination while you prepare your child for his/her injection.

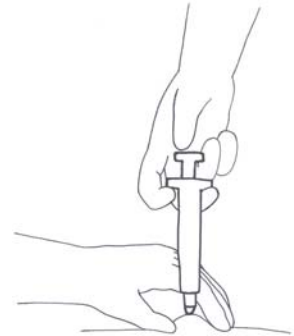


Giving the Injection

1. Choose a site for the injection. See diagram.
It is important to rotate the sites for injection.



2. Be sure the site is clean.
3. Pull the cap straight off the needle.
4. Hold the syringe in one hand, like a dart or pencil. Gently pinch up the skin at the chosen site with the other hand.
Remember not to touch the area you have cleaned.
5. Insert the needle quickly at a 90-degree angle. A 45-degree angle may be used for a very slim child who has little subcutaneous fat.
6. Inject all the medication in the syringe by pushing down on the plunger slowly.
7. Keep the needle in the skin for a few seconds.
8. Pull the needle straight out and apply pressure to the site with a clean dry gauze for approximately one minute. You may place a Band-Aid on the site if desired.
9. Do not recap the needle, in order to prevent sticking yourself. Dispose of your syringe and needle in a sharps container. All other supplies may be disposed of in the trash.
10. Record the date, time and site of injection.



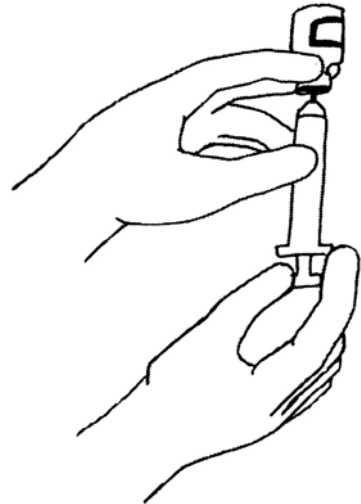
GIVING AN INTRAMUSCULAR INJECTION

Medication and dosage: _____

Supplies: _____

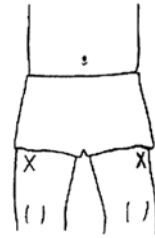
Drawing Up the Medication:

1. Wash your hands thoroughly.
2. Check that the medication is correct and not past the expiry date.
3. Clean the top of the bottle with an alcohol swab.
4. Attach and tighten needle on to the syringe.
5. Remove the needle protector. Do not let the sterile needle touch anything.
6. Pull back the plunger, drawing up the amount of air equal to the amount of medicine to be given.
7. Insert the needle into the rubber stopper on top of the bottle. Push the air into the bottle.
8. Turn the bottle and syringe upside down.
9. Pull down on the plunger, drawing up the correct amount of medicine. The end of the needle must be in the medicine to draw up medicine.
10. While the needle is in the medicine, look for large air bubbles. Remove them by pushing them back into the bottle. Recheck that you have the correct amount of medicine.



Giving the Injection:

1. Choose the site for the injection. A good site for children is the outer thigh, halfway down the thigh.
2. Clean the site with alcohol and let it dry.
3. Pull the cap straight off the needle.



4. Hold the syringe in one hand, like a dart or pencil. Support the skin at the chosen site with the other hand. You may have to pinch up the tissue if your child is thin. Remember not to touch the area you have cleaned.
5. Insert the needle quickly at a 90-degree angle.



6. Release the skin and check the location of your needle by pulling back on the plunger.
If there is blood on the hub of the needle, pull out and change the needle. Start again from Step 4.



7. Inject all the medication in the syringe by pushing down on the plunger slowly.
8. Pull the needle straight out and apply pressure to the site with a clean dry gauze for approximately one minute. You may place a Band-Aid on the site if desired.
9. Do not recap the needle in order to prevent sticking yourself. Dispose of your syringe and needle in a sharps container. All other supplies may be disposed of in the trash.
10. Record the date, time and site of injection.



Shopping list

For giving pills to a baby:

- Pill crusher
- Pill cutter
- Seven-day pill holder made of see-through plastic.

For giving oral syrup to a baby:

- Oral syringe.
Size and quantity needed _____
- Bottle adapter, to draw syrup into oral syringe.

For giving emergency hydrocortisone injections:

- Syringes _____ (size and quantity needed)
- Needles: _____ (size and quantity needed)

For giving growth hormone injections:

- Mixing equipment, if needed. _____

- Supplies for subcu injection _____
- Alcohol swabs
- Sharps disposal container.

For giving antidiuretic hormone, DDAVP®

For giving sex steroids

For checking blood glucose:

- Monitor: _____
- Strips: _____
- Lancets: _____
- Quick acting glucose: _____

- Other: Thermometer
 Pedialyte

Websites

The Magic Foundation: Panhypopituitarism
www.magicfoundation.org/www/docs/110/

The Pituitary Tumor Network Association
www.pituitary.org/

Medic Alert
www.medicalert.ca

Acknowledgements

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Graphics (appendix): Harry Toor

Thanks to Donna Williams for the parent's perspective on this challenging condition.

Thank you to the family and friends of Cameron McInroy whose generous gifts, given in his memory, have contributed to the publication of this booklet.

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BCCH1464
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