

Preventable Deaths: Panhypopituitarism and adrenal insufficiency. What you need to know

What is panhypopituitarism?

Your child has been diagnosed with a big scary sounding word, and all you can think is: 'What does this mean? and 'Why have I never heard of this?' Simply put, panhypopituitarism means that your child's pituitary gland does not function properly and as a result, your child is deficient in one or several hormones. Some children have congenital panhypopituitarism, meaning they are born with it. Others have acquired panhypopituitarism following an event such as head trauma, brain tumor surgery, or brain radiation. It is rare enough that is entirely possible, in fact, probable, that you will not initially know anyone else with this disorder.

What will my child need?

Although the diagnosis and condition can seem intimidating, it is very manageable once you understand what is needed. Unfortunately the condition cannot be cured or reversed, but again, it can be effectively managed. Your child will need to take medications to replace the missing hormones. These might include thyroid hormone, growth hormone, cortisol, and/or possibly others. Your doctor will go over these with you, as dosages vary from child to child. Most medications are taken orally, but growth hormone must be taken by daily injection. Your endocrinologist will work with you and your child to achieve the proper dosages and will guide you in how to administer any necessary medications.

Why is it sometimes life threatening? What is adrenal insufficiency?

Of the hormone deficiencies your child may have, the most critical is cortisol, also known as the 'stress hormone.' **Cortisol is essential for life**, and is therefore the central focus of this guide. In healthy people in whom the pituitary and adrenal glands are working normally, additional cortisol is automatically produced by the body in times of illness or serious injury. But in people with panhypopituitarism, or panpit or PHP for short, this does not occur. Therefore it is crucial that you know about and are comfortable with administering stress doses of cortisol to your child when needed. Your doctor will go over the specific guidelines, such as when to double or triple your child's cortisol dose.

The biggest issue faced by those with PHP is adrenal insufficiency brought on by illness or injury. Failure to address adrenal insufficiency can lead to serious consequences, including hypoglycemia (low blood sugar) and shock (very low blood pressure). If not properly treated, adrenal insufficiency is life threatening. It can occur with serious illness, or even with a minor illness when, for whatever reason, the oral stress doses of cortisol are not given (or are not absorbed by the body due vomiting or diarrhea) or are

given but are just simply not enough to counter the adrenal insufficiency. **It is important to know that not every illness leads to adrenal crisis in every child. But it is also important to grasp that sometimes something as common as an ear infection or a cold can lead to an adrenal crisis in a PHP child. Fortunately, there are signs and symptoms you can watch for, and steps you can take in the event they occur.**

What do I do if my child gets sick?

- 1. Give oral stress doses of cortisol per your doctor's previous instructions.**
- 2. Watch for signs or symptoms of adrenal insufficiency.** (Remember not all signs may be present so don't wait for them all to appear! See list below)
- 3. If you need to, give the 'emergency shot' (see below) and then get to the closest ER.** (There is no harm in giving the shot; not giving it may be life-threatening. If you are thinking about giving it, just do it!)
- 4. If you give the shot and go to the ER, take this pamphlet and the used bottle of SoluCortef with you to show what you have given, along with your child's other medications, or a current list of the medications and dosages.**
- 5. Tell ER personnel and any paramedics (if you go by ambulance) that your child is having an adrenal crisis.** It's not necessary that you try to explain panhypopituitarism at this point - that can sometimes lead to confusion and can instead be done once your child is stabilized. While they might not be familiar with panhypopituitarism, they ARE trained for what to do for an adrenal crisis.
- 6. Bring your child's endocrinologist into the loop to direct care at your earliest opportunity in this process.**

How will I know if it's an adrenal crisis?

Signs of an adrenal crisis vary from person to person. No two people are alike and no two sets of symptoms are alike, but there are some common signs. The most common ones are:

- lethargy, weakness, dizziness, or extreme listlessness (beyond normal tired)**
- glassy eyes**
- poor responsiveness, or in severe situations, complete unresponsiveness**
- nausea, vomiting**
- incoherent speech in older children**
- raspy breathing**
- poor skin color, very pale**
- seizure**

What is the Emergency Shot?

The emergency shot is a high dose of cortisol given into the muscle. As noted above, there are times when oral stress dosing is ineffective due to vomiting or diarrhea, or perhaps the oral stress dosing was started late, and an adrenal crisis occurs. In these instances an injection of SoluCortef is required. Many endocrinologists and PHP parents therefore call this the Emergency Shot. It must be carried at all times by the child and/or his parent or caretaker. It is different from growth hormone injections

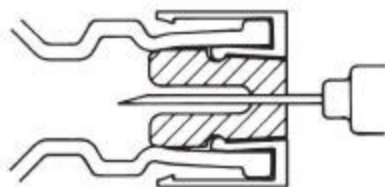
because it is given as an IM (intramuscular) injection, usually into the thigh, and because it goes straight into the muscle, it works quickly. Your child's doctor should provide you at the time of diagnosis with a prescription for several of these kits (ones for home, purse, school/daycare, any caregivers' house, etc) and should provide you with instructions on how to give it. If your child has been diagnosed with cortisol deficiency and your doctor has not prescribed it, please call him or her IMMEDIATELY and request it because it is absolutely critical that you have this. Make sure that the prescription is for the SoluCortef Act-o-Vial, which comes complete with the liquid diluent (vs just the powder that would be used in an IV line in the hospital.) Double check it before you leave the pharmacy. (There should be clear liquid in the top of the bottle separated from the powder in the bottom section by a red cork.) You will also need to be provided with the correct size syringes to give the shot. These syringes are larger than growth hormone syringes. Keep each SoluCortef vial with a syringe and alcohol swab so that everything is together when you need it. Be sure to check the expiration dates periodically and replace as needed.

DIRECTIONS FOR USING THE ACT-O-VIAL SYSTEM FOR SOLUCORTEF

Supplies Needed:

- SoluCortef emergency vial
- Alcohol Swab
- Syringe with needle

1. Press down on plastic activator to force fluid into the lower compartment of the vial.
2. Gently roll the vial to mix solution. Dissolve the powder completely.
3. Remove plastic tab covering center of stopper.
4. Sterilize top of stopper with an alcohol swab.
5. Insert needle squarely through center of stopper until tip is just visible (see the figure below). Turn vial upside down and withdraw appropriate dose
6. Discard any medicine remaining in the vial. (Note however that if there is another full dose left in the vial you can put the remainder in the fridge in case it needs to be repeated in the next 8-10 hours rather than waste it. You can then toss later if not used.)
7. Give injection in upper thigh using a dart-like motion, inserting the needle at a 90 degree angle.



Figure

But I hate needles!!

Nobody likes needles and no parent enjoys giving injections to his or her child. But with this diagnosis you must learn to work past that. It is strongly suggested that you first practice giving a SoluCortef shot to a piece of fruit such as an orange, so that you will feel comfortable if the time comes when you must give it. Yes, it 'wastes' a vial of medication, but it is an inexpensive medication, and more importantly, the moment your child is in crisis is not the best time to be learning how to do it. If you do ever have to give it, you will be very glad you practiced.

What else?

In addition to always carrying SoluCortef with your child, **your child should also wear a MedicAlert bracelet or necklace at all times.** (Note that ambulances in the United States do not carry SoluCortef, so it is necessary that a vial and syringe always be with your child.) Many parents use terms such as 'cortisol dependent' and 'needs stress dose steroids' on the MedicAlert bracelet, rather than 'panhypopituitarism', so that paramedics unfamiliar with that term will still know what is needed.

Note: If your child is school age, you will also need to have a plan in place with the school clinic for emergency situations. You can contact The MAGIC Foundation (see below) for more information and suggestions about how to set this up.

It is also suggested that you always keep a current printed list of your child's medications and dosages and the doctor's name and contact information. Give the list to any caregivers along with the emergency shot, and also keep it handy at home. This will save time and anxiety in an emergency situation.

Important Things to Remember:

- 1. You may never know for sure if it's the start of an adrenal crisis.**
- 2. You may have to trust your gut feelings or instincts.**
- 3. Don't wait for a list of symptoms to appear. There might only be one noticeable sign.**
- 4. The harm is not in giving the shot, it is in not giving it when needed.**
- 5. When in doubt, give the shot.**
- 6. Any fear of needles must be conquered. Your child's life may depend on it.**
- 7. What might be a minor illness in another child can lead to more serious illness in a PHP child if not treated properly.**
- 8. Every parent initially feels overwhelmed when first told of this diagnosis. But it truly is very manageable and will eventually become just another part of daily life. The key is to have knowledge about the disorder and be prepared for an emergency. Ask your doctor to explain anything you feel unsure of.**
- 9. This guide is NOT meant to replace a physician's advice, nor is it intended to be a comprehensive look at panhypopituitarism. It is meant to provide you with basic practical information regarding stress dosing of cortisol and in**

particular, the emergency injection of cortisol. Again, for further information, consult your doctor.

For additional resources and to network with other parents, you can contact:

MAGIC Foundation - www.magicfoundation.org or call at 708-383-0808.

Yahoo support group - go to Yahoo groups and type 'panhypopituitarism' in the search field and then follow the prompts to join the group.