MUSC Patient Handout
Homecare Handbook on Infantile Spasms and ACTH

Changing What’s Possible in Health Care.
AS AN ACADEMIC MEDICAL CENTER, MUSC offers benefits to the residents of South Carolina and beyond. The highly dedicated, professional team focuses on providing the most effective therapies available while also striving to develop new therapies within a highly active basic and clinical research program. These efforts are backed by facilities with the latest technology. This resource was developed to help patients understand more about their condition, treatment, or procedure. This handout is meant to be used as a guide to supplement your healthcare providers instructions. It is not intended to be used as a substitute for professional medical care. Only your physician can diagnose and treat a medical condition. Please consult your physician if you have any questions.

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Introduction

Your child has been diagnosed with Infantile Spasms and ACTH has been chosen as the medicine to try to stop them. This handbook will be a resource for you at home. It reviews all of the teaching done with you by the nurse and physician.

Infantile Spasms

Infantile Spasms consists of a specific seizure type which occurs in infants. The seizures or spasms are called flexor, extensor or mixed. The spasms usually last a few seconds and occur in clusters. An infant can have many clusters a day. Spasms usually occur during drowsiness and while awake.

The description of the spasms, your child’s neurological examination and history of development led to the consideration of the diagnosis of Infantile Spasms. The electroencephalogram (EEG) looks at brain wave patterns. The brain wave pattern in Infantile Spasms is called hypsarrhythmia and it helps confirm the diagnosis. The hypsarrhythmia pattern means the brain waves are very disorganized. The medication should stop the spasms and help the EEG pattern be less disorganized. The EEG may even return to normal.

Once the diagnosis is made, your infant will receive more tests to try to find out the cause of the Infantile Spasms. This is important to see if there is something else that needs treatment. It is also a factor in your infant’s care. A cause is found in over half of the cases of Infantile Spasms. In less than half, no cause is found. Most infants diagnosed with Infantile Spasms have some developmental problems that started before or because of the Infantile Spasms. These problems can range from mild to severe. A good response to the ACTH treatment may help improve the infant’s outcome.

About ACTH

ACTH (adrenocorticotropic hormone) is the treatment of choice for Infantile Spasms. It is a steroid. The goal of ACTH therapy is to stop the spasms and this usually occurs within one to two weeks of starting the medicine. ACTH is typically given over a twelve weeks period, but could be shorter or longer. The dose is decreased (tapered) slowly until it is stopped. Length of therapy depends upon the infant’s response.

The ACTH preparation recommended is called Acthar gel. The only way to give this medicine is by injection into the muscle. It is often frightening for parents to think about giving their baby a shot, but you will be given instruction and support to learn these skills.
ACTH SIDE EFFECTS

ACTH has many possible side effects, but these are not permanent. The side effects usually disappear or improve as the ACTH dose is tapered. The side effects need to be monitored and some, if they occur, require you to call the Neurology Office.

1. **Weight gain** – Infants receiving ACTH can develop rapid and significant weight gain. The face may appear chubby (also known as moon face) and a fat pad may develop between the shoulders. The weight increases due to fluid retention and increased appetite. These effects go away as the dose is tapered.

   **Things you can do to help:**
   a. Continue to feed your child normally, but as appetite increases, try to give half strength juice, fruit, rice/pasta (without a lot of salt or butter), etc.
   b. Check weight two times per week while on ACTH.
   c. If you are concerned about your child’s diet, we can arrange for our dietitian to talk with you.
   a. Blood pressure will be checked two times per week.

2. **High blood pressure** – This can occur because the body tends to hold more fluids while on this medication. If the blood pressure is high, additional medication may be prescribed to control it. Things you can do to help:
   a. Blood pressure will be checked two times per week.

3. **Irritability** – This is the hardest side effect for parents and families to cope with. The infant may cry inconsolably for hours and sleep may be disrupted.

   **Things you can do to help:**
   a. Maintain usual schedule for infant as possible.
   b. Use quieting activities to promote calm like soft music, giving your baby a bath, rocking in a chair, etc. Never shake your baby, if needed, put the baby in a safe place and walk away for a few minutes.
   c. Reducing daytime naps may help your infant sleep longer at night.
   d. Develop a schedule amongst family and friends to care for your infant to give you a break. You need time away to keep your spirits and energy up.
   e. Make sure the irritability is not mistaken for a wet diaper, hunger, illness, etc.
4. **Facial acne** – The acne will disappear as the ACTH is tapered.

**Things you can do to help:**
- Wash the face with mild soap and water only, acne medicines will not help at all.

5. **Increased susceptibility to infections** – ACTH suppresses (does not allow) the body’s ability to fight infection. It is important to avoid others who are ill, especially if they have the chickenpox or a fever blister. Watch for any signs or symptoms like: fever, runny nose, cough, diarrhea, vomiting, skin rash, increased sleepiness, etc. Immunizations should not be given during ACTH therapy. A common infection that can occur due to this side effect is called candida (or yeast infection). It will appear as white patches that will not rub off in the mouth or a red pimply rash in the diaper area. It is easily treated with medication.

**Things you can do to help:**
- If any signs or symptoms or illness noted, call pediatrician.
- Inform pediatrician that your child is on ACTH so that immunizations will not be given until therapy is completed.
- Observe mouth for white patches and diaper area for red pimply rash, and report to pediatrician if occurs.
- Keep your infant away from crowds and people who are ill. Ask family members to wash hands before handling your child.
- If infant is exposed to a person with chickenpox or any other illness call the pediatrician immediately.

6. **Stomach upset** – If this occurs, there are medicines which can easily treat this. Sometimes bleeding can occur in the gastrointestinal tract and the health care provider will need to be called.

**Things you can do to help:**
- If infant develops stomach upset, call pediatrician to obtain prescription for treatment.
- Observe stool for blood and if seen, call pediatrician.

7. **Nutritional effects** – ACTH can cause salt and water retention as well as potassium loss. It can cause the body to make too much sugar.

**Things you can do to help:**
- Avoid foods high in salt like: bologna, hot dogs, ham, bacon, sausage, canned spaghetti/ravioli, tomato soup, macaroni and cheese, potato chips, pretzels, corn chips, crackers with salt, catsup, mustard, pickles, relish, boullion or broth, tuna fish, all salt seasonings, olives, cheese, etc.
b. Include foods high in potassium like: grape juice, orange juice, bananas, dried fruits, potatoes, cooked cabbage, fresh winter squash, etc.

c. Check sugar in urine once per day, at the same time every day. Call Neurology for any positive 3+ glucose in urine.

WHEN TO CALL THE NEUROLOGY OFFICE OR PEDIATRICIAN

Call for any of the following:

1. Blood pressure greater than 120/80.
2. Temperature over 100.5°F
3. Signs or symptoms of an illness.
4. Thrush in the mouth or rash in diaper area.
6. Belly is puffy.
7. Greater than 3+ glucose in urine.
8. Infant does not want to eat.
9. Infant becomes unusually sleepy or lethargic.
10. Infant is exposed to chickenpox or another illness.
11. Seizures return or increase in number.

Please keep a record of the daily temperature, urine glucose and number of seizures noted. This record will be reviewed by the pediatrician and the neurology team. Please also write down any other information that would be helpful like blood pressures, weights, and side effects observed.

Follow-up with both the pediatrician and neurology is important. Your child will be seen more frequently until completion of the treatment. Further EEGs may be done also.

Please call the Neurology Office with any questions or concerns you may have at (843) 792-3307.

Pediatrician ________________________________

Phone Number ________________________________
CHECKING THE URINE FOR SUGAR

Supplies needed: Urine glucose sticks
   - Paper cup
   - Cotton to place in diaper to catch urine

1. Take wet cotton ball.
2. Squeeze urine from cotton ball into paper cup.
3. Dip urine glucose stick into urine and read after specified time noted on label for dipsticks.

GIVING THE ACTH INJECTION

Supplies: 23 gauge and 25 gauge, 1 inch needle
   - 1 cc tuberculin syringe
   - Alcohol pads
   - Acthar gel vial (80 units/cc)
   - Cotton balls
   - Needle box

*This job definitely takes 2 people. Decide before starting who will hold and who will give the injection.

A demonstration of how to draw up the medication will be shown to you.

1. ACTH is stored in the refrigerator. It may be taken out up to 15 minutes before the injection. The vial may be warmed by rolling it between the palms a few times (figure A). Do not shake the vial. All other supplies should be stored out of children’s reach.

2. After gathering all of the supplies, wash your hands with soap and water (figure B). Draw up the ACTH as previously demonstrated and according to the guide below.
   a. Pull air into the syringe equal to the amount of medicine to be given (figure C).
   b. Push 23 gauge needle through top of bottle. Push air into the bottle (figure D).
   c. Turn bottle upside down. Pull the amount of medicine that is to be given into the syringe (Figure E).
   d. Flicking the syringe with your thumb and index finger with the needle in the upright position will force the air bubbles to the top of the syringe (Figure F). The air can be pushed out by pushing the plunger up (Figure G). Remember to keep the bottle upside down all the time that you are drawing up the medication. After you have the
desired amount of medication, remove the needle. Change needle to 25 gauge for easier injection (Figure H).

3. Choose the injection site. See the diagram below to identify the injection site. It is best to use the thigh muscle in children who are not walking (Figure I).

4. Once you have located the area to inject, hold the muscle between your thumb and fingers, creating a bulge. Wipe the area with an alcohol pad in a circular motion (Figure J).

5. Take the syringe with the needle and remove the needle cap. Inject the needle quickly into the skin at a 90 degree angle (just as if you were throwing a dart) (Figure K). Pull back on the plunger slightly, if no blood comes back, you are in the muscle and may inject the medication. The gel is thick, inject it until all medicine is in and then withdraw the needle quickly (Figure L).

6. Apply gentle pressure for a few seconds with a cottonball (Figure M).

7. Dispose of the needle and syringe in the needle box. The needle box can be brought to the hospital for proper disposal once the treatment is over (Figure M).

8. Rotate sites each time to lessen the risk of injury to the muscle from repeated injections. Use the diagram (Figure N) as a guide.

Gigi Smith, RN, MSN, CPNP Division of Pediatric Neurology
Figure A
The vial of ACTH may be warmed by rolling it between the palms a few times.

Figure B
After gathering all of the supplies, wash your hands with soap and water.

Figure C
Pull air into the syringe equal to the amount of medicine to be given.
**Figure D**
Push 23 gauge needle through top of bottle. Push air into the bottle.

**Figure E**
Turn bottle upside down. Pull the amount of medicine that is to be given into the syringe.

**Figure F**
Flicking the syringe with your thumb and index finger with the needle in the upright position will force the air bubbles to the top of the syringe.
After you have the desired amount of medication, remove the needle. Change the needle to 25 gauge for easier injection.

The air can be pushed out by pushing the plunger up. Remember to keep the bottle upside down all the time that you are drawing up the medication.

Choose the injection site. See the diagram. It is best to use the thigh muscle in children who are not walking.
Figure J
Hold the muscle between your thumb and fingers, creating a bulge. Wipe the area with an alcohol pad in a circular motion.

Figure K
Inject the needle quickly into the skin at a 90 degree angle (just as if you were throwing a dart).

Figure L
Pull back on the plunger slightly. If no blood comes back, you are in the muscle and may inject the medication.

Figure M
Apply gentle pressure for a few seconds with a cottonball. Dispose of the needle and syringe in the needle box. The needle box can be brought to the hospital for proper disposal once the treatment is over.
Medical University of South Carolina

Patient’s Name: ______________________________________________________

You have been given a prescription for ACTH GEL _______________ Units per ML/CC.

Please give the average units, following the guides below, based on the prescription strength you have been given.

Special Instructions: _____________________________________________________________
____________________________________________________________________________
____________________________________________________________________________
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Figure N
Rotate sites each time to lessen the risk of injury to the muscle from repeated injections.
ACTH GEL 80 Units per ML/CC

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ACTH GEL 40 Units per ML/CC

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Pediatric Neurology ACTH Flow Sheet

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DOB: _________________________________________________________________

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