

## **Procedures/Risks: neurology**

### **Biopsy(muscle)**

*Procedure:* The biopsy requires a cut in the skin about 1 ½ inches long. Before a cut is made in the skin, the skin will be numbed with medicine through an injection. Lidocaine is a local anesthesia that dentists frequently use for dental procedures. At each biopsy, 3 small pieces of muscle (the size of an eraser on a pencil) will be removed. The biopsies will be performed at The Ohio State University. After the cut is made and the biopsy taken, the incision is closed with stitches that will need to be removed in 10-14 days.

*Risk:* Potential complications from the muscle biopsy procedure include: infection at the biopsy site which may require early stitch removal or possible antibiotic treatment.; bleeding at the site of the biopsy that could require additional stitches; and/or pain at the site of the biopsy.

Acetaminophen (also known as Tylenol<sup>®</sup>) will be offered every 4 hours as needed to control pain. There will be a scar at the site of the incision; usually this is small but occasionally some individuals have a tendency to develop bigger scars.

### **Electroencephalogram (EEG)**

*Procedure:* This study will use an electroencephalogram (EEG) to measure and record the electrical activity of your brain. In order to do this a number of electrical wires will be attached to electrodes (contact pads to which the wires are connected) placed on your scalp.

*Procedure:* An electroencephalogram (EEG) is a measurement of the electrical activity of the brain. Special sensors (electrodes) are attached to a person's head to pick up the natural electricity of the brain and measure how fast the "brain waves" are. No electricity is put into the brain or head; the EEG only measures what is there, like an electrocardiogram (ECG) measures the electricity in the heart.

*Risks:* Although there are no known adverse (bad or harmful) events associated with the EEG itself, it does require that the electrodes be glued to your head and scalp for the recording time. Your skin must be mildly roughened to make sure that the electrodes stay attached to your scalp during the recording.

### **Electromyography (EMG)**

*Procedure:* Electromyography (EMG) is a technique that is used to monitor muscle activity. The EMG is able to measure the electrical activity of muscle when they work (contract) and rest (relax). Electrode gel and alcohol will be used to scrub and clean your skin over the areas [name areas] where the muscle activity will be measured. Electrodes will be put on the muscles with double-sided adhesives. Tissue oxygen sensors will also be put over the muscles of interest.

*Risks:* There are no known risks to electromyography. All of the contact pads that are placed on your skin are marketed as hypoallergenic (unlikely to provoke an allergic reaction). However, there is still a possibility of skin irritation due to the experimental procedure. If you have sensitive skin you should alert the researchers.

## **Lumbar Puncture**

### *Procedure:*

Cerebrospinal fluid (CSF) will be collected via lumbar puncture by a qualified licensed healthcare professional. The lumbar puncture involves the insertion of a needle into the space around the spinal cord that is filled with CSF. Before this needle is inserted, an anesthetic will be injected to numb the area. Once the area is numb, the lumbar puncture needle will be inserted and up to 15 mL or 3 Teaspoons of CSF will be removed. Each lumbar puncture takes about 30 minutes. After the lumbar puncture, you may be asked to lie flat on your back in the exam room for about one hour.

### Post-Lumbar Puncture Follow-up Telephone Call (Visit 2.1)

This phone call will take place 48-hours after the Screening or Baseline Visit. This phone call may last about 10 minutes. The following procedures will be performed:

- Your medical status will be assessed.
- You will be asked about any side effects you may have experienced (like a headache).

### *Risks:*

#### Lumbar Puncture

Lumbar puncture is a standard procedure used in medical practice. When spinal fluid is removed during a lumbar puncture, the risks include headache, bleeding and pain at the site where the needle was put in, and infection. Pain during the lumbar puncture procedure will be prevented or minimized by using local anesthesia (lidocaine). Infection after a lumbar puncture is very rare, but serious, and would be treated with antibiotics.

About 1 out of 3 people who have a lumbar puncture develop a post-lumbar puncture headache. Headache can occur if the lining around the spinal fluid (dura) is torn and some of the fluid leaks out. Post-lumbar headaches are more common in females and in people less than 30 years old. This headache can be mild to severe. You may also have nausea, dizziness, and ringing in the ears.

If you develop a headache, you will need to lie down to reduce the headache pain and symptoms. Post-lumbar puncture headaches get worse when you are sitting or standing. Occasionally, the headache may be severe enough to interfere with your normal daily activities, such as going to work or school. If this happens, no payment will be made available to you for time missed from work or school or for others costs, such as paying for a babysitter.

If you get a headache, you should contact Stephen Kolb, M.D., Ph.D. at The Ohio State University Medical Center. Pain medication will be given to you, if needed. If the headache

lasts more than three days, a procedure called a blood patch may be performed. This procedure involves taking blood from your arm and injecting it in the same place where the spinal needle was put in during the lumbar puncture. The clotting of the blood in this space should stop further fluid leaking and stop the headache.

### **Measures of Muscle Power**

*Procedure:* A physical therapist will test your muscle strength [on your upper and/or lower limbs] using standard manual muscle testing and a computerized hand-held device; this testing is called myometry. Eight muscles on each side of the body including the shoulders, elbows, legs, hips and knees will be tested. The evaluation will take about 60 minutes.

Your muscle strength will be tested. To test your muscle strength, you will be asked to push or pull against the hand of the evaluator as much as possible for a few seconds. Next you will lie on an exam table and the evaluator will place a strap around your arm or leg and ask you to push or pull with your muscles against the strap as much as possible for a few seconds.

*Risks:* The risks and discomforts of the tests of muscle strength [and breathing] could include feeling tired, and muscle cramps.

### **MUNE and CMAP Measurements:**

*Procedure:* You will be asked to lie on your back or side during testing. Two electrodes (small disks through which electrical current flows) will be placed on your wrist and one electrode will be placed on the back of your hand. Several electrical shocks will be given on your wrist in order to measure the size of the muscle in your little finger. This will cause a slight tingling sensation in the wrist and finger that is somewhat similar to when you bump the “funny bone” in your elbow. This will take approximately 30 – 60 minutes.

*Risks:* Minor local discomfort, irritation and skin redness at the wrist may occur, where the electrodes are applied for the MUNE and CMAP. Also, there may be mild muscle soreness and fatigue after muscle testing that may last for one to two days. Like all other electrical devices and monitoring equipment connected to patients, this procedure carries the risk of stray leakage currents that under certain circumstances can result in an electrical injury, especially in patients in the intensive care setting. These complications are rare and are more likely to happen in certain populations including patients with pacemakers and other heart or cardiac devices. The testing procedures will be explained fully to you. Testing will be performed by testors experienced in all the procedures outlined to minimize physical discomfort.

### **Nerve conduction and/or muscle testing:**

*Procedure:* One or more small electrical shocks will be given to you [your child] on your [his/her] wrist in order to measure the size of the muscle in your little finger. This will cause a

slight tingling sensation in the wrist and finger that is somewhat similar to when someone bumps the “funny bone” in their elbow.

A test of how fast your nerves conduct their electrical signals called nerve conduction testing will be performed at each visit. This testing involves attaching electrodes (½ inch rectangular adhesive pads) over muscles in the hand. The electrodes taped over muscles in the hand will be connected to a machine that records the responses. You will lay in the position most comfortable for you [your child]. A nerve will be activated at the wrist and around the elbow by applying brief electrical shocks. This feels something like when you bang your “funny bone” at the elbow hard, and you get that tingly, electrical feeling down your arm. The total time for this testing is approximately 60 minutes.

Because this procedure can be somewhat uncomfortable or cause anxiety, a medication may be used for sedation (to make me more calm) during this testing. The medication is called diazepam, or better known by its drug brand name of Valium. This medication will be administered either inside the nose or by mouth. The dose of this drug will be carefully determined by your age and ability to cooperate. This medication may make you drowsy and minimize discomfort or anxiety during the studies, but you will not be asleep. To receive sedation, you must not have any food or drink for several hours prior to the procedure. A small clamp called a pulse oximeter will be placed on your toe to monitor heart rate and oxygen status throughout the procedure and for 30 minutes following the procedure. A blood pressure cuff will be placed on your upper arm or leg to obtain blood pressure readings prior to the start of the study and during the study. A nurse experienced in sedation will monitor you during the procedure and afterwards, and the doctor performing the electrical studies will also be there. Suction equipment and oxygen will be available at the bedside if needed. Prior to discharge from the hospital, you must have completely returned to a fully awake state and be back to normal. A nurse will monitor you after the procedure until these goals are achieved. [Your blood will be drawn prior to the nerve conduction testing.] The time required for the nerve conduction testing and monitoring is approximately 60 minutes.

*Risks:* There are no known risks associated with electrical stimulation of nerves. No complications, such as infections or nerve injury, have been reported from nerve [muscle] testing, but the test can be somewhat uncomfortable. Minor local discomfort, irritation and skin redness at the wrist may occur, where the electrodes are applied for the MUNE and CMAP. Also, there may be mild muscle soreness after muscle testing that may last for one to two days. The testing procedures will be explained fully to you. Testing will be performed by medical professionals experienced in all of the procedures that will be involved in this study to minimize physical discomfort.