

## **Argininosuccinic Acidemia (ASA)** (Argin-in-o-suc-sin-ic A-sid-EE-mia)

### **What is ASA?**

Argininosuccinic Acidemia (ASA) is a metabolic disorder. This means the body has a chemical imbalance. ASA is a condition in which the body is not able to process protein properly. People with this condition lack a certain enzyme. The enzyme is called argininosuccinic acid (ASA) lyase. The lack of this enzyme results in a build up of chemicals in the blood. These chemicals include ASA, citrulline, and ammonia. ASA is inherited. This means it is present at birth.

### **What type of problems occurs with ASA?**

A baby who has this condition may refuse to eat and have vomiting. If untreated, this condition can lead to seizures and coma. Early diagnosis and treatment will help prevent these problems.

### **What is the chance my baby will have ASA?**

This condition occurs in less than 1 in every 100,000 births. Babies born with this condition have a changed gene from each parent. A person who has one changed gene is called a carrier. A person who is a carrier does not have symptoms. If both parents are carriers, either parent can pass on the changed gene to their baby. If both parents pass on the changed gene, the baby will have the condition. If both parents are carriers, for each pregnancy:

- There is a 25% chance that the baby will be born with this condition.
- There is a 50% chance that the baby will be a carrier for this condition.
- There is a 25% chance that the baby will not be born with this condition and will not be a carrier.

### **What is the treatment of ASA?**

The treatment for this condition is usually a low protein diet. A supplement called arginine might be needed. Your baby's metabolic doctor will help you make sure that your baby gets the right diet and medical care. Routine illness, like the common cold, can make this condition worse. So be sure to contact your baby's doctor whenever your baby gets sick. Sometimes a liver transplant is needed.

### **Where in Virginia can I take my baby for care?**

There are three different treatment centers in Virginia. The names of the centers and the metabolic doctor are listed below.

- Virginia Commonwealth University, Richmond, Virginia, Dr. Arti Pandya
- Eastern Virginia Medical School, Norfolk, Virginia, Dr. Virginia Proud
- University of Virginia, Charlottesville, Virginia, Dr. Bill Wilson

If you want to know more about this condition, please contact Virginia Newborn Screening Services, Virginia Department of Health. The telephone number is 804-864-7714 or 804-864-7715. The Web site is <http://www.vahealth.org/genetics>.



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