



## Newborn Screening FACT Sheet

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# Argininosuccinic Acidemia (ASA)

### What is ASA?

ASA is a type of amino acid disorder. People with this condition can't remove ammonia from the body. Ammonia is a harmful substance. It is made when the body breaks down protein and amino acids for use by the body.

### What Causes ASA?

ASA is a "urea cycle disorder" (UCD). ASA happens when an enzyme called "argininosuccinic acid lyase" (ASAL) is missing or not working. Enzymes help start chemical reactions in the body. Ammonia builds up in the blood when there is a problem with the ASAL enzyme. Too much ammonia in the blood can cause brain damage. It can also cause death if not treated.

### What Symptoms or Problems Occur with ASA?

*[Symptoms are something out of the ordinary that a parent notices.]*

There are two kinds of ASA. The severe form starts in babies. They are healthy when born, but soon show symptoms of high ammonia levels. The milder form of ASA starts in childhood.

Some of the first symptoms of high ammonia are:

- poor appetite
- too much sleepiness or no energy
- irritable mood
- vomiting

If not treated, high ammonia can cause:

- muscle weakness
- breathing problems
- problems staying warm
- seizures
- swelling of the brain
- coma, sometimes leading to death

The milder form can also cause intellectual disability, seizures, a large liver, and skin and hair problems.

### What Is the Treatment for ASA?

The following treatments are often used for babies and children with ASA:

**1. Low-protein diet and/or special medical foods and formula** – The best treatment is a very low-protein diet. There are medical foods such as special low-protein flours, noodles, and rice available. A dietitian will make a food plan for your child. Dietitians know what the right foods to eat are. Your child will need to eat a low-protein diet for life. The doctor or dietitian may give your baby a special formula that has the right nutrients and amino acids.

**2. Medication** – The doctor might prescribe arginine supplements for your child. Other medicines may be used to prevent high ammonia.

**3. Blood tests** – Regular blood tests will check your child's amino acid and ammonia levels.

### Things to Remember

Children with high ammonia often need to be treated in the hospital. Call your doctor right away if your child has any of the following:

- loss of appetite
- low energy or too much sleepiness
- vomiting
- fever
- bad headache
- infection or illness
- behavior or personality changes (such as crying for no reason)
- problems walking or balancing