Argininosuccinic Acidemia (ASA) & Citrullinemia Type I (CIT-I) General Overview

Q. What are ASA and CIT-I?

A. ASA and CIT-I are two similar disorders that affect the way the body processes protein. They are treatable, but can cause life-threatening illness.

Q. What happens to ammonia in a child with ASA or CIT-I?

A. When the body breaks down protein to make energy, ammonia is made as a waste product and then turned into urea (part of the urine). In a child with ASA or CIT-I, ammonia cannot be turned into urea because certain enzymes (chemical substances) do not work properly. This results in dangerous levels of ammonia that build up in the blood and are toxic to the brain and the rest of the nervous system.

Q. What is the treatment for ASA and CIT-I?

A. Both ASA and CIT-I are treated by a special formula and foods that are low in protein. Many children also need medication to help them eliminate ammonia. Treatment must begin shortly after birth and is life-long. Some doctors may also prescribe dietary supplements. People with ASA or CIT-I require treatment through a specialty genetics clinic with experience in treating these disorders.

Q. Why would a child have ASA or CIT-I?

A. ASA and CIT-I are inherited disorders. They result when a baby receives a double-dose of the same non-working gene (one from each parent). For more information about this, contact your health care provider or a genetic counselor.

Q. How common is ASA and CIT-I?

A. About one in every 128,000 babies is born with ASA or CIT-I. However, parents who have a child with ASA or CIT-I each carry one copy of the same non-working gene. That means, with each pregnancy, there is a one in four chance of the child having ASA or CIT-I.

For more information about ASA and CIT-I, please see the Disorders section of our website: www.doh.wa.gov/nbs.

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