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Climb™

Supporting those affected by Inherited Metabolic Disorders

# Argininosuccinic Aciduria

Argininosuccinic Aciduria is a Urea Cycle Disorder, in which the body's ability to manage dietary protein is impaired. When the body digests protein it is broken down into small molecules known as amino acids. Excess amounts of these amino acids are converted into a toxic substance known as ammonia. In the liver, the ammonia is converted to urea and excreted in the urine. In Urea Cycle Disorders one of the six enzymes does not function properly and ammonia is not removed from the blood stream. In Argininosuccinic Aciduria there is a deficiency or absence of the enzyme argininosuccinate lyase (ASL) which is an important part of the urea cycle. This leads to an accumulation of the amino acid, Argininosuccinic Acid (hence the name) and may lead to a build up of ammonia (and its related product glutamine) in the body, which cause the symptoms of the disorder.

### Synonyms

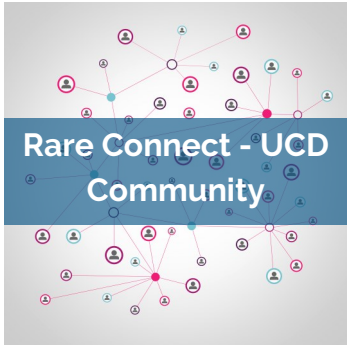
Alternative names for this condition are:

- Argininosuccinase Deficiency
- Argininosuccinate Lyase Deficiency
- Argininosuccinic Acidaemia
- ASA
- ASL

Further information about this condition is available from Climb.

### Disclaimer

Please read our disclaimer and information on data protection.



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