Need to talk? Call us 0845 2412173 Monday to Friday 9am to 5pm

Climb™

Supporting those affected by Inherited Metabolic Disorders

Isovaleric Acidaemia

Isovaleric Acidaemia (IVA) is a relatively rare metabolic disorder that belongs to a group of disorders known as the Organic Acidaemias. It is caused by a deficiency of an enzyme known as Isovaleryl CoA Dehydrogenase (IVD) which is needed by the body to break down the amino acid leucine into isovaleric acid. If the enzyme is deficient, isovaleric acid builds up and becomes toxic to the body. Isovaleric Acid causes a characteristic odour that occurs when individuals have an acute metabolic crisis. Symptoms of a crisis include, irritability, breathing problems, lethargy and vomiting.

IVA is now routinely tested for shortly after birth as part of the Newborn Bloodspot Screening Programme. If IVA is diagnosed, treatment is started immediately. Newborn screening and early management means that most children with IVA are now able to live normal, healthy lives. Without lifelong treatment and management, severe and life-threatening symptoms can develop, including brain damage, seizures or coma.

Synonyms

Alternative names for this condition are:

- IVA
- Isovaleric Acid CoA Dehydrogenase Deficiency
- Isovaleryl CoA Carboxylase Deficiency
- IVD Deficiency

Further information about this condition is available from Climb.

Disclaimer

Please read our disclaimer and information on data protection.

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Telephone: **0845 241 2173** Freephone: **0800 652 3181**
Email: contact@climb.org.uk Website: www.climb.org.uk