Climb™

Supporting those affected by Inherited Metabolic Disorders

Crigler-Najjar Syndrome Type 1

Crigler-Najjar Syndrome is characterised by an absence of a liver enzyme called bilirubin-UGT which is needed to allow the body to excrete bilirubin, a substance created when red blood cells are broken down. The enzyme is needed to perform a chemical reaction to change a toxic form of bilirubin (unconjugated bilirubin) into a harmless form (conjugated bilirubin) in order for it to be excreted by the body. People with Crigler-Najjar Syndrome Type 1 are unable to convert the bilirubin and therefore have a build-up of the toxic form in their blood (unconjugated hyperbilirubinaemia). There are two forms of Crigler-Najjar Syndrome, type 1 where there is a complete absence of the enzyme, and type 2; a milder form caused by a partial deficiency.

Symptoms present shortly after birth with jaundice and severely high levels of bilirubin in the blood. Severe hyperbilirubinaemia can lead to a life-threatening condition called kernicterus which can cause severe damage to the brain and central nervous system.

Synonyms

Alternative names for this condition are:

- CN1
- Familial Nonhemolytic Unconjugated Hyperbilirubinemia
- Hereditary Unconjugated Hyperbilirubinemia
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Further information about this condition is available from Climb.

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