

Supporting those affected by Inherited Metabolic Disorders

## Leigh Syndrome

Patients with Leigh Disease usually develop symptoms in the first months of life, although in some cases onset of symptoms may not be until later in childhood or even (occasionally) in adulthood. Affected individuals characteristically suffer degeneration of the nervous system as a result of failure of energy production in the mitochondria of brain cells. Mitochondria are the cell's 'batteries' and produce energy in the form of ATP. Energy is produced by burning sugars or fats within the mitochondria. The final pathway for generating energy for fuels is known as the respiratory chain which is located within the wall of the mitochondria. Problems within the respiratory chain can lead to a lack of energy and lead to a build up of the things that would usually be burnt. For example from the carbohydrates there will be an accumulation of lactic acid (lactic acidosis). Due to all parts of the body requiring energy to function, Leigh's Disease is a multisystem disorder.

## **Synonyms**

Alternative names for this condition are:

- Leigh Disease
- Necrotising Encephalomyelopathy (of Leigh)
- Subacute Necrotising Encephalomyelopathy
- SNF

Further information about this condition is available from Climb.



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