

**DISEASE:**  
**Mitochondrial DNA-associated Leigh syndrome**

<b>NAME:</b>	Mitochondrial DNA-associated Leigh syndrome
<b>DESCRIPTION:</b>	Maternally inherited Leigh syndrome is a rare subtype of Leigh syndrome (see this term) characterized clinically by encephalopathy, lactic acidosis, seizures, cardiomyopathy, respiratory disorders and developmental delay, with onset in infancy or early childhood, and resulting from maternally-inherited mutations in mitochondrial DNA.
<b>ORPHACODE:</b>	255210
<b>SYNOMYS:</b>	MILS Maternally-inherited Leigh disease Maternally-inherited infantile subacute necrotizing encephalopathy mtDNA-associated Leigh syndrome
<b>XREF(S):</b>	<a href="#">Orphanet</a> <a href="#">ICD-10</a> <a href="#">OMIM</a>

<b>ANALYTE(S):</b>	MT-ATP6 MT-ND1 MT-ND2 MT-ND3 MT-ND4 MT-ND5 MT-ND6 MT-TL1 MT-TK MT-TW MT-TV
<b>CREATED:</b>	13 May 2019 - 01:02
<b>CHANGED:</b>	22 Jun 2023 - 16:14

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## RELATED CONTENT

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### Related Genetic Tests

- [Hepatorenal disorders \(gene panel\)](#)
- [Leigh / NARP Syndrome](#)
- [Mitochondrial complex III deficiency / Bjornstad syndrome / Gracile syndrome](#)

### Related Laboratories

- [Centre de Génétique Médicale UCL](#)
- [Centrum Medische Genetica - UZ Brussel VUB](#)

### Related Analytes

- [mitochondrially encoded ATP synthase membrane subunit 6](#)
- [mitochondrially encoded NADH:ubiquinone oxidoreductase core subunit 1](#)
- [mitochondrially encoded NADH:ubiquinone oxidoreductase core subunit 2](#)
- [mitochondrially encoded NADH:ubiquinone oxidoreductase core subunit 3](#)
- [mitochondrially encoded NADH:ubiquinone oxidoreductase core subunit 4](#)
- [mitochondrially encoded NADH:ubiquinone oxidoreductase core subunit 5](#)
- [mitochondrially encoded NADH:ubiquinone oxidoreductase core subunit 6](#)
- [mitochondrially encoded tRNA-Lys \(AAA/G\)](#)
- [mitochondrially encoded tRNA-Leu \(UUA/G\) 1](#)
- [mitochondrially encoded tRNA-Val \(GUN\)](#)
- [mitochondrially encoded tRNA-Trp \(UGA/G\)](#)

## Related Gene Panels

- [Leigh syndrome \(mtDNA / 37 genes\) - VUB](#)

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