Morava et al., 2006 criteria for mitochondrial disorder†

**I. Clinical signs and symptoms**
(1 point/symptom, max 4 points)

I.A. Muscular presentation (max 2 points)
___ Ophthalmoplegia (2 points)
___ Facies myopathica
___ **Exercise intolerance**
___ Muscle weakness
___ Rhabdomyolysis
___ Abnormal EMG

I.B. CNS presentation (max 2 points)
___ **Developmental delay**
___ **Loss of skills**
___ Stoke-like episodes
___ Migraine
___ **Seizures**
___ Myoclonus
___ Cortical blindness
___ Pyramidal signs
___ Extrapyramidal signs
___ Brain stem involvement

I.C. Multisystem disease (max 3 points)
___ Hematology
___ **GI tract**
___ **Endocrine/growth**
___ Heart
___ Kidney
___ Vision
___ Hearing
___ Neuropathy
___ Recurrent/familial

**Please consult your physician for help with interpretation**

**II. Metabolic/imaging studies**
(1 point/symptom, max 4 points)
___ **Elevated lactate (2 points)**
___ Elevated lactate/pyruvate ratio
___ **Elevated alanine (2 points)**
___ Elevated CSF lactate (2 points)
___ Elevated CSF protein
___ Elevated CSF alanine (2 points)
___ Urinary tricarbon acid excretion (2 points)
___ Ethylmalonic aciduria
___ Stroke-like picture MRI
___ Leigh syndrome/MRI (2 points)
___ Elevated lactate/MRS

**III. Morphology (muscle biopsy)**
(1 point/symptom, max 4 points)
___ Ragged red/blue fibers (4 points)
___ Cox-negative fibers (4 points)
___ Reduced COX staining (4 points)
___ Reduced SDH staining
___ SDH positive blood vessels (2 points)
___ Abnormal mitochondrial/EM (2 points)

**Score:**
1: mitochondrial disorder unlikely
2 to 4: possible mitochondrial disorder
5 to 7: probable mitochondrial disorder
8 to 12: definite mitochondrial disorder

†Reference:
Morava, E., L. van den Heuvel, F. Hol, M.C. de Vries, M. Hogeveen, R.J. Rodenburg, et al. (2006). Mitochondrial disease criteria: diagnostic applications in children. *Neurology, 67*, 1823-6.