

# Newcastle Mitochondrial Centre

**At a glance guidelines:**

## **Stroke-Like Episodes in Adult Mitochondrial Disease**

For full guideline visit:

<http://www.newcastle-mitochondria.com/service/patient-care-guidelines/>

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There are many different forms of mitochondrial disease, varying greatly in their genetic basis, clinical presentation, progression and prognosis. We recommend referral to a specialist mitochondrial centre for appropriate counselling and guidance ([www.mitochondrialncg.nhs.uk](http://www.mitochondrialncg.nhs.uk)).

Stroke-like episodes are a rare but serious feature of some forms of mitochondrial disease, often resulting in neuronal loss and accrued disability. We therefore recommend the following:

1. Mitochondrial stroke-like episodes are characterised by gradual evolution of headache, encephalopathy, seizures and focal deficits. Not all features are uniformly present. Stroke-like episodes should not be confused with the rapid loss of function typical of strokes of an atherosclerotic or thrombo-embolic aetiology.
2. Always consider other causes. Even in patients with established mitochondrial disease other treatable causes of encephalopathy, seizures or neurological deficit should be considered. This may necessitate urgent imaging and/or CSF studies.
3. Seizures should be treated actively from the outset, in order to minimize subsequent cerebral damage related to frequent or intractable seizures.
4. Non-convulsive status epilepticus should be actively excluded in patients with focal deficits or features of encephalopathy.

5. Sodium valproate should be avoided where possible and especially in patients who might carry POLG mutations due to potential fulminant hepatotoxicity.
6. There are no available drugs with proven efficacy in the treatment of stroke-like episodes. Benefit from L-arginine has been proposed but further trials are needed. Focus should be directed to seizure control and supportive care.
7. Potential precipitants (eg infection, dehydration, metabolic derangements) should be sought and treated.
8. Comorbidities (eg cardiomyopathy, ileus) should be considered, particularly in the ITU patient.
9. Discussion with a specialist mitochondrial centre is advised at the earliest opportunity. Advice regarding investigation and management will be available. Confirmation/exclusion of common causative mutations may be available with a rapid turn-around for those patients not already diagnosed.