



wellcome  
centre  
mitochondrial  
research

**NHS Highly Specialised  
Services for Rare  
Mitochondrial Disorders**

**At a glance guidelines:**

# **Peri-operative Care in Children with Mitochondrial Disease**

For full guideline visit:

<http://www.mitochondrialdisease.nhs.uk/professional-area/care-guidelines>

## **First published March 2018**

Mitochondrial diseases are a diverse group of neurometabolic conditions presenting with a plethora of clinical features and at varying ages. The genetics, progression and prognosis are equally variable. We recommend referral to a specialist mitochondrial centre in all cases [www.mitochondrialdisease.nhs.uk](http://www.mitochondrialdisease.nhs.uk)

Children with mitochondrial disease risk decompensation during times of metabolic stress. All anaesthetic agents can potentially suppress the mitochondrial respiratory chain. There is limited evidence for the use of general anaesthesia in children with mitochondrial disease, however adverse effects appear rare. Children with more severe multi-system disease appear most at risk. Careful planning and preparation can help to reduce peri-operative complications, as advised below.

### **1. Pre-operative planning**

- 1.1 Liaison with a mitochondrial specialist prior to surgery can minimise risks.
- 1.2 Prior knowledge of genotype, phenotype including multi-system involvement will help preoperative planning
- 1.3 Baseline bloods including FBC, U&Es, LFTs, calcium, phosphate, glucose, CK and lactate should be considered
  - 1.3.1 FGF-21 is currently being reviewed as a clinical marker and consideration should be given to analysis prior to surgery if a mitochondrial disease is suspected
- 1.4 ECG and ECHO should be considered in those children with mtDNA mutations or nDNA mutations with cardiac associations
  - 1.4.1 Cardiac rhythm disturbances or cardiomyopathy should be discussed with a paediatric cardiologist prior to anaesthesia
- 1.5 FVC may be helpful in those children able to cooperate to exclude respiratory muscle / diaphragmatic weakness

- 1.6 Swallow assessment should be considered if there are concerns about dysphagia
- 1.7 Treatment of constipation is advised pre-operatively; post-operative paralytic ileus can occur (especially in patients with m.3243A>G or MNGIE)
- 1.8 All relevant information should be discussed with the Paediatric anaesthetist prior to surgery
- 1.9 Consideration for booking a HDU or PICU bed should be made
- 1.10 Muscle or skin biopsy may be required for further analysis, or research purposes; liaise with the mitochondrial specialist and surgical team whether or not this is possible when the child is anaesthetised.

## **2 Pre-operative Management**

- 2.1 Fasting should be minimised; IV fluids should be started early. Normoglycaemia should be maintained
- 2.2 Important regular medications such as anti-epileptic drugs should be administered; if this is not possible then an alternative preparation / formulation should be given e.g. IV instead of oral
- 2.3 Children with diabetes should be managed as per the local hospital protocol for patients with diabetes undergoing surgery

## **3 Anaesthesia**

- 3.1 There is no robust evidence to support malignant hyperthermia as a complication of GA in children with mitochondrial disease
- 3.2 Propofol appears safe for induction, provided it is not a rapid infusion of a large bolus
- 3.3 Prolonged propofol use (i.e. maintenance anaesthesia) may increase the risk of lactic acidosis

- 3.4 Etomidate, ketamine and barbituates are complex I inhibitors but there is no robust evidence to suggest that these anaesthetic agents are harmful
- 3.5 Muscle relaxants should be avoided in those with respiratory muscle weakness unless absolutely necessary
- 3.6 Lactate buffers e.g. Ringer's solution should be avoided when prescribing IV fluid therapy

#### **4 Post-operative Management**

- 4.1 Complications are most likely to occur in the post-operative period
- 4.2 Minimise fasting; if enteral nutrition is likely to be delayed, consider TPN early to ensure calorific requirements are met
- 4.3 Continue regular medications as soon as able; alternative formulations may be required
- 4.4 If muscle / skin biopsies were taken, contact the relevant mitochondrial laboratory
- 4.5 Routine blood monitoring is not required unless there are clinical concerns
- 4.6 Attention should be given to appropriate analgesia
- 4.7 Constipation and / or paralytic ileus is not uncommon post-operatively, particularly in patients with m.3243A>G or MNGIE; ***see Constipation guideline for further management***
- 4.8 Inform all professionals involved in the child's care about the surgical procedure and complications

## References

Footitt EJ, Sinha MD, Raiman JAJ, Dhawan A, Moganasundram S and Champion MP **Mitochondrial disorders and general anaesthesia: a case series and review** *Br J Anaesth* 2008; 100(4): 436-41

Driessen J, Willems S, Dercksen S, Giele J, Van der Staak F, Smeitink J. **Anaesthesia-related morbidity and mortality after surgery for muscle biopsy in children with mitochondrial defects.** *Pediatr Anesth* 2007; 17: 16-21

Morgan PG, Hoppel CL, Sedensky MM. **Mitochondrial defects and anaesthetic sensitivity.** *Anaesthesiology* 2002; 96: 1268-70

Wallace JJ, Perndt H, Skinner F et al. **Anaesthesia and mitochondrial disease.** *Pediatr Anaesth* 1998; 8: 249-54

Kam PCA, Cardone D. Propofol infusion syndrome. *Anaesthesia* 2007; 62 (7)

Driessen JJ. **Neuromuscular and mitochondrial disorders: what is relevant to the anaesthesiologist?** *Curr Opin Anaesth* 2008; 21: 350-355