At a glance guidelines:

Anaesthesia & Peri-Operative Care in Adult Patients:

Screening and Subsequent Management
There are many different forms of mitochondrial disease, varying greatly in their clinical features. The spectrum and severity of multi-system involvement helps guide the level of caution and preparation required in the pre and peri-operative period. All patients due to undergo significant surgery or a general anaesthetic should have their diagnosis highlighted to the anaesthetist. We recommend discussion with a specialist mitochondrial centre at the earliest opportunity and review of the full guideline (www.newcastle-mitochondria.com).

Most mitochondrial disorders have the potential for complications to arise in the peri-operative period. This also applies to asymptomatic carriers. We therefore recommend the following:

1. **Pre-operative preparations:**
   1.1. **Planning and Preparation:** planning and liaison with a mitochondrial specialist will minimise risks. Routine screening programmes (see existing guidelines) ensure knowledge of multi-system involvement.
   1.2. **Bloods:** FBC, U&Es, LFTS, Ca\(^{2+}\), Mg\(^{2+}\), glucose, HbA1c, CK and lactate
   1.3. **Cardiac:** ECG and ECHO
   1.4. **Respiratory:** FVC (erect and supine) to exclude respiratory muscle/diaphragmatic weakness.
   1.5. **Bowel Care:** ensure bowel care optimised pre-op. Consider AXR pre-op for later comparison. Risk of chronic constipation or post-op paralytic ileus (esp m.3243A>G or MNGIE)

2. **Pre-operative Management:**
   2.1. **Minimise Fasting:** fluids and calorific intake should be maintained in the pre-operative period. 50g carbohydrate in liquid form 2 hours pre-op (non-diabetic patients).
2.2. **Important drugs:** (eg AEDs) should be administered or replaced with alternative formulations

2.3. **Diabetes:** mitochondrial diabetes may be managed in the usual way (metformin avoided)

### 3. Anaesthesia:

3.1. Good evidence is lacking for benefit or harm specific to any anaesthetic agent. Muscle relaxants are best avoided in those with significant respiratory muscle weakness unless absolutely necessary.

3.2. There is no credible evidence for an increased risk of malignant hyperthermia syndrome.

3.3. Propofol appears safe for induction. Prolonged use for maintenance of anaesthesia may risk exacerbation of lactic acidosis.

3.4. Intravenous fluids: avoid lactate buffers (eg Ringer’s Solution)

3.5. Risk appears proportionate to severity of comorbidities (esp. cardiorespiratory disease).

### 4. Post-operative Management:

4.1. **See Pre-operative Care:** minimise fasting, administer drugs, usual post-op diabetic care.

4.2. **Bowel Care:** constipation or paralytic ileus is common in the post-operative period (esp m.3243A>G or MNGIE). Dilated loops on AXR should be compared to pre-op AXR. Discussion with a mitochondrial specialist advised ASAP and surgical intervention avoided (see published GI guidelines).

4.3. **Critical Care:** consider pre-planned admission for post-op monitoring