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 (www.mitochondrialncg.nhs.uk or see appendix for international centres).

Most have the potential for cardiac involvement and this may develop in the absence of symptoms. We therefore recommend the following:

1. All patients should be offered an ECG and trans-thoracic echocardiogram following initial diagnosis.
2. The ECG and echocardiogram should be repeated annually. This interval should only be extended after discussion with a specialist.
3. All patients due to undergo significant surgery or a general anaesthetic should have their diagnosis highlighted to the anaesthetist. Recent cardiac investigations (as above) should be available for review.
4. Treatment with conventional agents should be initiated with any evidence of hypertrophic remodelling regardless of symptomatic status.
5. Patients fulfilling conventional guidelines (European Society of Cardiology or American College of Cardiology / American Heart Association) for implantation of permanent pacemakers should be offered this therapy without delay, due to the unpredictable nature of progression.
Appendix A: Screening / management algorithm

Mitochondrial Disease: Cardiology Guidance

Stable for 3 years
Yearly ECG
Yearly ECHO

Abnormal
Conduction defect, High risk, or paroxysmal symptoms
Cardiology Opinion
24 hour tape
Evidence of CHB or TFB
Permanent Pacemaker

Evidence for SVT or WFW
Consider Ablation

Left Ventricular Hypertrophy, Dilated or Hypertrophic Cardiomyopathy
Cardiology Opinion
Start treatment with ACEi/ARB and Beta blockers
Progression despite Rx

Consider Increase or Change in Treatment

Key:
ACEi angiotensin converting enzyme inhibitor
ARB angiotensin II receptor blocker
CHB complete heart block
TFB trifascicular block
SVT supra-ventricular tachycardia
WPW Wolff-Parkinson-White syndrome