

Medications Contraindicated for Patients with Mitochondrial Disease

This patient has a possible or confirmed mitochondrial disease. The mitochondria are organelles within all cells of the body aside from red blood cells and serve to generate ATP to meet cellular needs. As many medications have an inhibitory effect (direct or indirect) on normal mitochondrial function (i.e. damage the mitochondrion itself, or impair ATP production), certain medications may cause a patient’s mitochondrial disease manifestations to deteriorate. We have listed the most commonly used medications that are most likely to have an adverse effect on baseline function when used in a mitochondrial disease setting. Other non-listed medications, may also have an adverse effect but are not listed because of one of two reasons: the medication is used rarely in regular practice (eg AZT), or the potential adverse effect has not been verified and may be small in comparison to the potential benefits. When starting a new medication, unlisted in the table, in a mitochondrial disease patient, contact our clinic if you have any doubt about the relative safety.

Pharmacologic Category	Medication	Mechanism of Mitochondrial Toxicity
Anticonvulsants	Valproate	Uncoupler, inhibition of fatty acid oxidation, acyl CoA, carnitine depletion, toxic metabolite 2, 4 diene valproate.
Anesthetic Agents	Propofol	Inhibition of electron transport chain
Analgesics	ASA (for high doses and at any dose for children)	Uncoupler leading to decreased ATP generation efficiency.
Antibiotics	Gentamicin	Most toxic other aminoglycosides less so
Antibiotics	Linezolid	Inhibits ribosomal 50s subunit inhibiting mitochondrial protein synthesis

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