

Table 2. Points of attention regarding drug prescription in patients with a mitochondrial disease

Specific drug / situation / disease causing gene	Points of attention
Specific drug / disease causing gene	
Valproic acid (sodium valproate)	Should be used only in exceptional circumstances. Should absolutely not be given to patients with mutations in a gene called POLG or to patients with symptoms suspicious for POLG disease. Should not be used in patients with liver disease.
Aminoglycosides (antibiotics)	In case of specific mitochondrial DNA mutations (12S rRNA) these antibiotics may cause hearing loss. In emergency situations aminoglycosides could be used while the benefits of the drugs are more important then. If long-term treatment is necessary one should screen for these mutations and / or switch to another antibiotic regime.
Neuromuscular blocking drugs (used for anaesthesia)	In patients with muscle disease these drugs should preferably not be used or, if necessary, under strict monitoring.
Specific situation	
General anaesthesia and surgery	The time of fasting before surgery should be as short as possible. During prolonged anaesthesia fluid and caloric intake should be guaranteed by glucose infusion, unless the patient is on a ketogenic diet. Side effects may develop when medication is used for
Duration of treatment	a longer period. It must be assessed for each individual patient whether the need for long-term treatment outweighs the possible side effects. Kidneys remove certain medicines from the blood. If
Kidney failure	the kidneys do not work properly, too much medicine remains in the blood. This may cause more side effects. Dosage and / or dosing frequency should be adjusted for these medicines. Patients with a mitochondrial disease may have an
High lactic acid in blood	increased blood acidity due to high lactic acid. In that case, drugs that can make the blood acidic should preferably not be used or, if necessary, under regular monitoring of blood values.