

L-Arginine Fact Sheet for Physicians

- Mitochondrial myopathy, encephalopathy, lactic acidosis and stroke-like episodes (MELAS) is a mitochondrial electron transport chain disorder. POLG (polymerase gamma) mutations lead to mitochondrial DNA depletion/deletions. Patients with both MELAS and POLG can have cerebral infarcts. These stroke-like episodes may be due to the impairment of arterial vasodilatation related to abundance of intracellular reactive oxygen species that inactivate nitric oxide.
- L-arginine has been used chronically and acutely to prevent and treat stroke-like episodes, with promising results.
- Studies show that long term oral arginine is associated with normalization of brachial artery vasodilatation responsiveness and decreased frequency and severity of stroke-like episodes
- Studies show that administration of IV arginine to MELAS and POLG patients with acute stroke-like episodes is associated with more rapid symptomatic improvement and increases of plasma nitric oxide and cGMP
- Our recommendations:

	Oral arginine	IV arginine
dosage	start at 1000mg BID with food	for acute stroke-like episode – 3.3g/m ² rounded to the nearest 0.5g) in D5W 100mL IV Q8H x 3doses infuse over 30 minutes
monitoring	Monitor plasma amino acids to keep arginine trough levels >80 umol/L. Begin testing one month after arginine therapy is initiated. If no adjustments are needed, test yearly with other bloodwork.	-monitor electrolytes -monitor plasma amino acids at 24 and at 48 hours after the start of arginine infusion #1 (goal is for plasma arginine to remain at 200 umol/L) **patient should still be assessed as r-TPA candidates if they meet eligibility criteria

Our contact information

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After hours/holidays: 604-875-4111; ask for the neurologist on call

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Page 2 of 2

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