**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:

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<tr>
<th>Oral arginine</th>
<th>IV arginine</th>
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<tr>
<td>dosage start at 1000mg BID with food</td>
<td>for acute stroke-like episode—3.3g/m² rounded to the nearest 0.5g in D5W 100mL IV Q8H x 3 doses infuse over 30 minutes</td>
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| 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**

The Adult Metabolic Diseases Clinic  
Vancouver General Hospital  
Monday-Friday 8:00-4:00 604-875-5965  
After hours/holidays: 604-875-4111; ask for the neurologist on call
References


