Cardiomiopatia mitochondrial se beneficia com L-arginina: aumenta da função do ciclo de Krebs com aumento da função miocárdica como bomba

Abnormal myocardial energy-production state in mitochondrial cardiomyopathy and acute response to L-arginine infusion. C-11 acetate kinetics revealed by positron emission tomography.
Source
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Abstract
BACKGROUND:
Cardiomyopathy is a life-threatening condition in patients with mitochondrial myopathy, encephalopathy, lactic acidosis and stroke-like episodes (known as MELAS). However, no effective therapy has been available until now. In the present study cardiac energetics and acute effects of L-arginine (Arg) were evaluated in MELAS patients.

METHODS AND RESULTS:
The 6 patients with MELAS (M-group) and 6 volunteers (C-group) underwent dynamic C-11 acetate positron emission tomography (PET) imaging. TCA-cycle metabolic rate (k(mono)), myocardial efficiency (double product (DP)/k(mono)), and myocardial blood flow (MBF) were determined before and after L-Arg administration. Baseline k(mono) showed a lower value in the M-group than in the C-group (0.051±0.013 vs 0.070±0.019 min(-1), P=0.055). On the other hand, baseline DP/k(mono) was significantly greater in the M-group (1.69±5.9 vs 0.95±1.2×10(5), P=0.004). After L-Arg administration, 4 patients showed significant elevation of k(mono). No relationship was observed between the distribution of k(mono) elevation and the increase in MBF.

CONCLUSIONS:
The TCA cycle metabolic rate is markedly suppressed in MELAS patients, indicating a shift in energy production to the anaerobic pathway, leading to a paradoxical increase in myocardial efficiency. L-Arg can enhance TCA-cycle metabolism, regardless of its vasodilatation effect, and can be used as a treatment for patients with mitochondrial cardiomyopathy.

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– C-11 Acetate Kinetics Revealed by Positron Emission Tomography –
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