# eTable 2. Excluded studies after full-text review

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| **Author ID** | **Title** | **Reason for exclusion** |
| Abraham, Abraham & Khanna, 20101 | Arginine extravasation leading to skin necrosis | Patients did not meet criteria for genetically confirmed diagnosis |
| Ebrahimi-Fakhari et al. 20152 | Recurrent Stroke-Like Episodes in FBXL4-Associated Early-Onset Mitochondrial Encephalomyopathy | Insufficient detail provided to ascertain if L-arginine was used for the acute or prophylactic treatment of SLEs |
| El-Hattab et al. 20123 | Restoration of impaired nitric oxide production in MELAS syndrome with citrulline and arginine supplementation | No L-arginine intervention |
| El-Hattab et al. 20164 | Impaired nitric oxide production in children with MELAS syndrome and the effect of arginine and citrulline supplementation | L-arginine was not used for the acute or prophylactic treatment of SLEs |
| Goto & Momoi 20045 | Treatment for mitochondrial diseases | No L-arginine intervention |
| Ikawa, Povalkoc & Koga, 20186 | Arginine therapy in mitochondrial myopathy, encephalopathy, lactic acidosis, and stroke-like episodes | Duplicated data of previously published (and included) article (Koga et al. 2018) |
| Koga et al. 20067 | Endothelial dysfunction in MELAS improved by L-arginine supplementation | Duplicated data of previously published (and included) article (Koga et al. 2005) |
| Koga et al. 20078 | MELAS and L-arginine therapy | Duplicated data of previously published (and included) article (Koga et al. 2005) |
| Koga et al. 20089 | L-arginine therapy on MELAS | Duplicated data of previously published and included) article (Koga et al. 2005) |
| Liu et al. 201910 | Mitochondrial A3243G mutation causes mitochondrial encephalomyopathy in a Chinese patient | No L-arginine intervention |
| Moutaouakil et al. 200911 | L-arginine efficiency in MELAS syndrome. A case report  | Unable to translate to English (French) |
| Murakami & Ono 201712 | MELAS: Mitochondrial encephalomyopathy, lactic acidosis and SLEs | No L-arginine intervention |
| Parikh et al. 200913 | A modern approach to the treatment of mitochondrial disease | No L-arginine intervention |
| Parikh et al. 201714 | Patient care standards for primary mitochondrial disease: A consensus statement from the mitochondrial medicine society | No L-arginine intervention |
| Rodan et al. 201515 | L-arginine Affects Aerobic Capacity and Muscle Metabolism in MELAS (Mitochondrial Encephalomyopathy, Lactic Acidosis and Stroke-Like Episodes) Syndrome | Patients did not meet criteria of SLEs |
| Rodan et al. 202016 | L-arginine effects on cerebrovascular reactivity, perfusion and neurovascular coupling in MELAS (mitochondrial encephalomyopathy with lactic acidosis and stroke-like episodes) syndrome | L-arginine was not used for the acute or prophylactic treatment of SLEs |
| Song et al. 201917 | Twenty-one-year follow-up of variable onset MELAS syndrome with heteroplasmic mt3243A>G mtDNA mutation: A case report | Patients did not meet criteria of SLEs |
| Sudo, Sano, & Kawamura 201418 | Determination of the critical time point for efficacy of L-arginine infusion therapy in a case of MELAS with frequent SLEs | Unable to translate to English (Japanese)  |
| Toribe 200719 | Usefulness of L-arginine infusion for status epilepticus in mitochondrial myopathy, encephalopathy, lactic acidosis, and SLEs | Unable to translate to English (Japanese) |
| Yatsuga et al. 201220 | MELAS: a nationwide prospective cohort study of 96 patients in Japan | No L-arginine intervention |

Abbreviations: SLE, stroke-like episode

NB: Only 1 patient (patient 1) was eligible for inclusion in Suzuki et al. 201721 (patient 2 and 3 excluded: no L-arginine intervention);

2 patients (1 and 3) included in Renard & Ion 202022 (patient 2 excluded: no L-arginine intervention).

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