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(71) Applicants *(for all designated States except US)*: **MCGILL UNIVERSITY** [CA/CA]; 845 Sherbrooke Street West, Montréal, Québec H3A 1B1 (CA). **CHRONOGEN** [CA/CA]; 1550 Metcalfe Street, Suite 502, Montréal, Québec H3Z 1X6 (CA).

(72) Inventors; and

(75) Inventors/Applicants *(for US only)*: **HEKIMI, Siegfried** [CA/CA]; 4335 Powell, Montréal, Québec H4P 1E5 (CA). **HIHI, Abdelmadjid** [DZ/CA]; 2006 Marlowe, Montréal, Québec H4A 3L5 (CA). **LEVAVASSEUR, Françoise** [FR/FR]; 20, rue du Moulin, F-50450 Gavray (FR). **SHOUBRIDGE, Eric** [CA/CA]; 222 avenue Metcalfe, Westmount, Québec H3Z 2H9 (CA). **GAO, Yuan** [CN/US]; 14 South Street, Unit 61, Danbury, CT 06810 (US). **PAQUET, Michel** [CA/CA]; 3200 avenue Mont Royal, Montréal, Québec H1Y 3K9 (CA). **BÉNARD, Claire** [CA/CA]; 3601 Ste-Famille #1004, Montréal, Québec H2X 2L6 (CA).

(74) Agents: **OGILVY RENAULT** et al.; Suite 1600, 1981 McGill College Avenue, Montreal, Québec H3A 2Y3 (CA).

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(54) Title: PHENOTYPIC EFFECTS OF UBIQUINONE DEFICIENCIES AND METHODS OF SCREENING THEREOF

(57) Abstract: The present invention relates to a method of screening for a compound allowing survival of clk1 homozygous mutant embryos; a method of screening for a compound suitable for rescue of mutant phenotype of mclk1 homozygous cell line; a method of screening for a compound suitable for partial or complete functional replacement of endogenous ubiquinone; a method for screening a compound capable of inhibiting activity of clk-1 and/or other processes required to make ubiquinone from demethoxyubiquinone; a non-ubiquinone-producer mouse; a DNA construct, which comprises an alteration of mclk1; a non-ubiquinone-producer ES cell line; a coq-3 mutant subject non-ubiquinone producer; a method of screening for a compound suitable for complete or partial functional ubiquinone or demethoxyubiquinone replacement; a method for reducing and/or increasing ubiquinone level in a multicellular subject; a method of screening for a genetic suppressor of clk-1; and a method of screening for a genetic suppressor of coq-3.

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