

表 2 (Compounds under Development for Treatment of Mitochondrial Disease in US)

	Generic Name	Trade Name	Designation Date	Designation	Pharmacology	Contact Company	Country
1	Levocarnitine	Carnitor	1997/4/7	Treatment of zidovudine-induced mitochondrial myopathy.	Supplement therapy	Sigma-Tau Pharmaceuticals, Inc.	US
2	Ubiquinone	Ubi-Q-Gel	1999/12/14	Treatment of mitochondrial cytopathies.	Anti-oxidant	Gel-Tec, Division of Tishcon Corp.	US
3	2',3',5'-Tri-O-Acetyluridine	n/a (RG2133)	2003/1/13	Treatment of mitochondrial disease	Cytoprotective agent	Repligen Corporation	US
4	Alpha-Tocopherol Quinone	n/a	2006/3/28	Treatment of inherited mitochondrial respiratory chain diseases	Anti-oxidant	Penwest Pharmaceuticals	US
5	Idebenone	RAXONE/ CATENA	2009/5/22	Treatment of mitochondrial myopathy, encephalopathy, lactic acidosis with stroke-like episodes syndrome (MELAS)	Anti-oxidant CoQ10 analog	Santhera Pharmaceuticals Limited	CH
6	Alpha-Tocotrienol Quinone	n/a (EPI-743)	2010/10/21	Treatment of inherited mitochondrial respiratory chain diseases.	Anti-oxidant	Edison pharmaceuticals, Inc.	US
7	Recombinant Thymidine Phosphorylase Encapsulated With Autologous Erythrocytes	n/a	2010/12/13	Treatment of mitochondrial neurogastrointestinal encephalomyopathy due to thymidine	Enzyme replacement therapy	St. George's University of London	GB
8	Cytochrome C, Flavin Mononucleotide And Thiamin Diphosphate	n/a	2011/6/17	Treatment of mitochondrial disorders	Supplement therapy	NBI Pharmaceuticals, Inc	US
9	modified recombinant mitochondrial transcription factor A (TFAM) containing the mitochondrial transduction domain	n/a	2012/8/20	Treatment of inherited mitochondrial respiratory chain diseases	Activator of mitochondrial transcription as well as a participant in mitochondrial genome	Gencia Corporation	US