

Online Table 1. mtDNA sequence data of 17 primate species online

<u>Species name</u>	<u>GenBank accession number</u>
Homo sapiens	NC_011137
<i>Cebus albifron</i>	NC_002763
<i>Gorilla gorilla</i>	NC_011120
<i>Hylobates lar</i>	NC_002082
<i>Lemur catta</i>	NC_004025
<i>Macaca mulatta</i>	NC_005943
<i>Macaca sylvanus</i>	NC_002764
<i>Nycticebus coucang</i>	NC_002765
<i>Pan paniscus</i>	NC_001644
<i>Pan troglodytes</i>	NC_001643
<i>Papio hamadryas</i>	NC_001992
<i>Pongo pygmaeus</i>	NC_001646
<i>Pongo pygmaeus abelii</i>	NC_002083
<i>Tarsius bancanus</i>	NC_002811
<i>Mus musculus</i>	NC_006914.1
<i>Bos taurus</i>	HM045018.1
<i>Xenopus laevis</i>	NC_001573.1

Online Table 2 mtDNA mutations in three Chinese families with Leber's hereditary optic neuropathy.

Gene	Position	Replacement	CRS ^a	WZ510	WZ511	WZ512	AA change	AA Position	Conservation (H/B/M/X)	Previously reported ^b
D-loop	73	A to G	A	G	G	G				Yes
	152	T to C	T	C						Yes
	228	G to T	G	T						Yes
	263	A to G	A	G	G	G				Yes
	310	T to CTC/TC	T	CTC	TC	TC				Yes
	489	T to C	T	C	C	C				Yes
	16066	A to G	A			G				Yes
	16093	T to C	T	C	C					Yes
	16129	G to A	G	A	A					Yes
	16223	C to T	C	T	T	T				Yes
	16249	T to C	T	C						Yes
	16311	T to C	T		C	C				Yes
	16362	T to C	T	C						Yes
12S rRNA	709	G to A	G		A	A			G/A/A/-	Yes
	750	A to G	A	G	G	G			A/A/A/-	Yes
	1438	A to G	A	G	G	G			A/A/A/G	Yes
16S rRNA	2706	A to G	A	G	G	G			A/G/A/A	Yes
	3010	G to A	G	A					G/G/A/A	Yes
	3168					Ins C				No
	3206	C to T	C	T					C/A/T/A	Yes
<i>ND1</i>	3483	G to A	G	A						Yes
	3866	T to C	T	C	C	C	Ile-Thr	187	I/I/I	Yes
	4035	A to G	A			G				No
<i>ND2</i>	4140	C to T	C		T	T				Yes
	4769	A to G	A	G	G	G				Yes
	4883	C to T	C	T						Yes
	5178	C to A	C	A			Leu-Met	237	L/T/T/T	Yes
	5466	A to G	A	G			Thr-Ala	333	T/S/S/S	Yes
<i>COI</i>	6357	C to T	C		T					Yes

	6915	G to A	G			A	Val-Met	338	V/M/M/M	No
	7028	C to T	C	T	T	T				Yes
	7250	A to G	A			G				Yes
	7295	A to C	A	C						No
<i>ATP8</i>	8414	C to T	C	T			Leu-Phe	17	L/F/M/W	Yes
	8473	T to C	T	C						Yes
<i>ATP6</i>	8701	A to G	A	G	G	G	Thr-Ala	59	T/S/L/Q	Yes
	8793	T to C	T		C	C				Yes
	8821	T to C	T		G		Ser-Ala	99	S/S/S/S	Yes
	8856	G to A	G		A	A				Yes
	8860	A to G	A	G	G	G	Thr-Ala	112	T/A/A/T	Yes
<i>CO3</i>	9540	T to C	T	C	C	C				Yes
	9966	G to A	G		A		Val-Ile	254	V/V/V/V	Yes
<i>ND3</i>	10197	G to A	G	A			Ala-Thr	47	A/A/A/M	Yes
	10398	A to G	A	G	G	G	Thr-Ala	114	T/T/T/A	Yes
	10400	C to T	C	T	T	T				Yes
<i>ND4L</i>	10646	G to A	G		A	A				Yes
<i>ND4</i>	10873	T to C	T	C	C	C				Yes
	11257	C to T	C		T					Yes
	11719	G to A	G	A	A	A				Yes
<i>ND5</i>	12549	C to T	C		T	T				Yes
	12705	C to T	C	T	T	T				Yes
	13135	G to A	G		A		Ala-Thr	267	A/T/T/M	Yes
	13152	A to G	A		G	G				Yes
	13401	T to C	T	C						Yes
	13561	C to A	C			A	Leu-Met	409	I/I/I	No
<i>ND6</i>	14502	T to C	T		C	C	Ile-Val	58	I/I/I	Yes
	14668	C to T	C	T						Yes
<i>Cytb</i>	14766	C to T	C	T	T	T	Thr-Ile	7	T/S/T/S	Yes
	14783	T to C	T	C	C	C				Yes
	14979	T to C	T	C			Ile-Thr	78	I/I/L/L	Yes
	15040	C to T	C		T					Yes
	15043	G to A	G	A	A					Yes

15071	T to C	T		C		Tyr-His	109	Y/F/F/Y	Yes
15218	A to G	A		G	G	Thr-Ala	158	T/T/T/N	Yes
15301	G to A	G	A	A	A				Yes
15326	A to G	A	G	G	G	Thr-Ala	194	T/M/I/I	Yes
15412	T to C	T	C						Yes

^aConservation of amino acid for polypeptides or nucleotide for RNAs in homo sapiens (H), bovine (B), mouse (M) and *Xenopus laevis* (X).

^bSee the online mitochondrial genome database <http://www.mitomap.org> and <http://www.genpat.uu.se/mtDB>

Supplemental Figure 1

I187T



Homo sapiens	STLITTTQ E HLWLLLPSWPLAMMWFISTLAETNRTPPFDLAE G ESELVSGF
Gorilla gorilla	STLIMTQ E HLWLLLPTWPLAMMWFISTLAETNRTPPFDLAE G ESELVSGF
Cebus albifrons	QSLITTTQ E QSWLLLPSWPLTMMWFISTLAETNRAPFDL T E G ESELVSGF
Hylobates lar	STLITTTQ E HIWLLLPTWPLAMMWFISTLAETNRTPPFDL T E G ESELVSGF
Lemur catta	STLITTTQ E YLWLIIPSWPLAMMWFISTLAETNRAPFDL T E G ESELVSGF
Macaca mulatta	N T LITTTQ E HLWLLLPSWPLAMMWF T STLAETNRTPPFDL M E G ESELVSGF
Macaca sylvanus	N T LITTTQ E HLWLLLPSWPLAMMWF T STLAETNRTPPFDL M E G ESELVSGF
Nycticebus coucang	STLIKTQ E HTWLLLPSWPLAMMWFISTLAETNRAPFDL T E G ESELVSGF
Pan paniscus	STLITTTQ E HLWLILPTWPLAMMWFISTLAETNRTPPFDL T E G ESELVSGF
Pan troglodytes	STLVTTQ E HLWLILPTWPLAMMWFISTLAETNRTPPFDL T E G ESELVSGF
Papio hamadryas	N M LITTTQ E HLWLILPSWPLAMMWF T STLAETNRTPPFDL M E G ESELVSGF
Pongo pygmaeus abelii	SALITTTQ E HSWLLLPSWPLALMWFISTLAETNRAPFDL T E G ESELVSGF
Pongo pygmaeus	STLITPQ E NSWLLLPSWPLALMWFISTLAETNRAPFDL T E G ESELVSGF
Tarsius bancanus	TKLIMTQ E YSWLIAPSWPLAMMWF T STLAETNRAPFDL T E G ESELVSGF
Bos taurus	STLITTTQ E QMWLILPAWPLAMMWFISTLAETNRAPFDL T E G ESELVSGF
Mus musculus	QTLITTTQ E HMWLLLPAWPMAMMWFISTLAETNRAPFDL T E G ESELVSGF
Xenopus laevis	TTLMTTQ E QMWLIIPGWPMAMWYISTLAETNRAPFDL T E G ESELVSGF

Supplemental Figure 2

