Table S1: Characteristics of included studies

Number	Author Year	Geography	Number of LHON Patients	Number of LHON Patients with G11778A	Proportion of G11778A in LHON	Age of G11778A LHON Patients	Gender Ratio in G11778A LHON Patients (M/F)
1	Qiao et al. 2015 [1]	China	4	4	/	mean: 40.3 ± 30.7 median:37 range: 12-75	1/3
2	Lu et al. 2017 [2]	China	8	8	/	mean: 45.8 median: 36.5 range: 27-88	4/4
3	Wan et al. 2016 [3]	China	9	9	/	mean: 19.2 ± 11.6 median: 17 range: 9-46	7/2
4	Du et al. 2011 [4]	China	18	10	55.6%	/	/

5	Cui et al. 2013 [5]	China	50	38	76%	/	/
6	Jiang et al. 2015 [6]	China	1281	453	35.4%	/	108/12 (analyzed)
7	Ji et al. 2008 [7]	China	619	619	/	/	436/183
8	A.A. Sadun et al. 2006 [8]*	Brazil	75	2	/	patient 1: 12 patient 2: 44	2/0
9	A.A. Sadun et al. 2002 [9]	Brazil	20	20	/	mean: 46.8 ± 17.2	17/3
10	A.A. Sadun et al. 2003 [10]	Brazil	20	20	/	/	17/3
11	F. Sadun et al. 2004 [11]	Brazil	20	20	/	mean: 47 ± 17 range: 14-84	17/3
12	Ramos Cdo et al. 2009 [12]	Brazil/ Italy	71	45	63.4%	/	/
13	Guy et al. 2014 [13]**	US	45	1	/	19	1/0
14	Feuer et al. 2016 [14]	US	5	5	/	mean: 44.2	4/1

						range:35-55	
15	Guy et al. 2017 [15]	US	14	14	/	mean: 33.8 median: 30 range: 19-56	10/4
16	Lam et al. 2014 [16]	US	44	44	/	mean: 32.1±13 range: 10-61	35/9
17	Ueda et al. 2017 [17]	Japan	44	38	86.4%	/	/
18	Mashima et al. 2017 [18]	Japan	61	61	/	/	56/5
19	Mishra et al. 2017 [19]	India	40	11	27.5%	mean: 19.6 ± 8.8 median:16 range: 11-42	11/0
20	Khan et al. 2017 [20]	India	146	146	/	/	112/34
21	Majander et al. 2017 [21]	UK	27	13	48.1%	/	10/3
22	Dimitriadis et al. 2014 [22]	Germany	253	167	66%	/	13/3 (late onset)

23	Spruijt et al. 2006 [23]	Netherland	351	145	41.3%	/	124/21
24	Kirkman et al. 2009 [24]	UK/ Netherlands /Germany	196	132	67.3%	/	/
25	Klopstock et al. 2011 [25]	UK/Canada /Germany	85	57	67.1%	/	/
26	Kim et al. 2003 [26]	Korea	82	46	56.1%	/	/
27	Chuenkongkaew et al. 2005 [27]	Thailand	62	62	/	mean: 37.3 ± 12.9 range: 8-68	47/15
28	Gowri et al. 2020 [28]	India	55	23	41.8%	/	/
29	Tonagel et al. 2021 [29]	Germany	33	18	54.5%	/	7/0 (analyzed)
30	Yu-Wai-Man et al. 2020 [30]	France/Germany/Italy/	37	37	/	/	29/8

		UK/US					
31	Ishikawa et al. 2021 [31]	Japan	57	54	94.7%	mean: 35.3 ± 17.9 median: 32.3 range: 11.2-75	49/5
32	Marotta et al. 2020 [32]	Australia	76	52	68.4%	mean: 26.4	40/12
33	Li et al. 2020 [33]	China	132	84	63.6%	mean: 28.7 ± 16.0 median: 25.5 range: 0.8-66	56/28
34	Ahn et al. 2020 [34]	Korea	64	38	59.4%	/	10/2 (early onset)
35	Poincenot et al. 2020 [35]	Global	1512	1044	69.0%	/	795/249
36	Yang et al. 2020 [36]	China	9	9	/	mean: 19.2 ± 11.6	7/2

						median: 17 range: 9-46	
37	Cui et al. 2019 [37]	China	73	73	/	/	49/6 (analyzed)
38	Dokrungkoon et al. 2019 [38]	Thailand	13	10	76.9%	/	/
39	Liu et al. 2019 [39]	China	80	80	/	/	68/12
40	Newman et al. 2021 [40]	France/Germany/Italy/UK/US	39	39	/	mean: 36.8 ± 15.4 range: 15-69	31/7 (analyzed)
41	Zhao et al. 2020 [41]	China	33	29	87.9%	/	/

LHON = Leber hereditary optic neuropathy.

* In the study of Sadun et al. 2006, 2 out of 75 individuals converted from G11778A carriers to LHON-affected patients.

** In the study of Guy et al. 2014, 1 out of 45 individuals converted from G11778A carrier to LHON-affected patient.

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