Online-Only Supplemental Materials

Appendix 1: Electronic search strategy, coding-scheme for the systematic review and data analysis

The search strategy was designed by a clinical investigator with relevant domain expertise in neurology (AAT).

We searched MEDLINE for English-language articles, using the following strategies with the following components: (1) defining the clinical syndrome (i.e., ataxia), (2) ocular motor or vestibular features, and (3) quantitative assessments). We did not expressly search for hereditary ataxia syndromes, as this was found to result in omitting most relevant studies due to lacking referral to the genetic background. We also performed a manual search of reference lists from eligible articles, and contacted corresponding authors where necessary. We did not seek to identify research abstracts from meeting proceedings or unpublished studies.

MEDLINE Search *(accessed via PubMed at www.ncbi.nlm.nih.gov/pubmed)*

(ataxia OR ataxic OR (gait AND impairment)) AND ((eye movements) OR (ocular motor) OR oculomotor OR vestibular OR saccade OR (smooth pursuit) OR (vestibulo-ocular reflex) OR VOR OR optokinetic OR nystagmus OR gaze OR head impulse OR caloric) AND (quantitative OR recording OR recordings OR quantified OR measured)

Search Results

Our search identified 819 unique citations, of which 624 (76.2%) were excluded at the abstract level (Figure 1, main manuscript). A record was excluded only if two scorers (PG, AAT) recommended exclusion (detailed list of predefined reasons for exclusion shown below). We did not demand concordance on reason for abstract exclusion, but, among those abstracts with concordant reasons for exclusion (63.3%, n=395), the distribution was as follows: 44.2% had no data on human subjects with ataxia; 6.6% were not reporting on the assessment of oculomotor /vestibular features; 6.1% were not reporting on quantitative measurements; 5.8% had no original data and 0.6% were not in English.

We further examine 195 full manuscripts (this included 4 articles identified by hand-search). After initial screening, there were a total of 10 disagreements about study inclusion for the two reviewers (PG and AAT, kappa=0.89 [95% CI: 0.85 – 0.94]). These differences were resolved by discussion. Overall agreement on reason for exclusion was 93.1%. We demanded concordance on reason for full-text exclusion and resolved differences by discussion.

At the end of our full-text review, 702 were excluded and 117 were considered eligible (Figure 1, main manuscript). These eligible studies represented 14.3% of the total (n=819). Among all full-text manuscripts excluded (40.0%), the distribution of reason for exclusion was as follows: 25.1% were not reporting on quantitative measurements; 9.7% had no data on human subjects with ataxia; 2.6% were not reporting on the assessment of oculomotor /vestibular features; 2.1% were not in English and 0.5% had no original data.

Coding schema for abstract and full-text reviews

All gathered literature was subject to title/abstract screening by two independent reviewers (PG and AAT). Full-text screening was then applied to all citations considered eligible or possibly eligible by at least one reviewer. Two independent reviewers (PG and AAT) determined whether full-text manuscripts were eligible and, if not, provided a reason for exclusion. Differences were resolved by discussion and consensus. AAT completed a hand search of the reference lists of selected articles for additional citations. For citations identified by hand search, the full process was repeated iteratively until no additional manuscripts were found for inclusion. We calculated inter-rater agreement on full-text inclusion using Cohen's kappa.[1] A formal review protocol was not registered or posted.

Abstract Review Coding Rules
1) Coding status options are "Yes", "No", "Maybe". We will review full text of "Yes" and "Maybe". The purpose of "Yes" vs. "Maybe" is to look at kappa values agreement on "Yes" vs. "Maybe".
2) Err on the side of "Maybe" if there is doubt about a "No"; this is more conservative.
3) If there is only a title, exclude it only if you feel confident; otherwise code it as "Maybe".
4) Each "No" or "Maybe" should be coded with a reason for exclusion.
5) Reasons for exclusion are listed below 0-5. Go through them in order from 0 to 5 for each abstract, coding the first reason for exclusion only, not multiple reasons for exclusion. Only code "0" for "not English" if you are sure it is "not English".
6) Two independent raters will code reason for exclusion, but we will not mandate agreement on exclusion reason at the abstract level.
7) Occasionally an abstract seems inappropriate for another reason. In such cases, code as "other". There should be few "other" codings.

Abstract Reasons for Exclusion

0	not English	manuscript is not in English
1	no data	review paper; no original patient data
2	not ataxia	No data on human subjects with ataxia
3	not oculomotor / vestibular	not reporting on the assessment of oculomotor and/or vestibular features
4	not quantitative	not reporting on quantitative oculomotor and/or vestibular measurements
5	other	any other reason abstract is not included

Full-Text Review Coding Rules

1) Coding status options are “Yes” or “No”.
2) Each "No" should be coded with a reason for exclusion.
3) Reasons for exclusion are listed below 0-4. Go through them in order from 0 to 4 for each full text, coding the first reason for exclusion only, not multiple reasons for exclusion.
4) Two independent raters will code reason for exclusion, and we will mandate agreement on exclusion reason at the full text level.
5) Coding differences will be adjudicated or consensus will be developed through dialogue.

Full-Text Reasons for Exclusion

0	not English	manuscript is not in English
1	no data	review paper; no original patient data
2	not ataxia	No data on human subjects with ataxia
3	not oculomotor / vestibular	not reporting on the assessment of oculomotor and/or vestibular features
4	not quantitative	not reporting on quantitative oculomotor and/or vestibular measurements

Appendix 2: quality assessment for studies reporting on oculomotor findings in ataxia

Two independent reviewers (PG and AAT) rated all included studies with regards to their quality in reporting oculomotor testing in ataxia patients. Discrepancies were resolved by discussion. Based on the eight items listed below an overall quality rating (high, moderate, low) was assigned. "High quality" was defined as having high quality on items 1, 3-7 and a low risk-of-bias for item 8. Item 2 was not considered since, depending on the study design a control group may not be required (e.g. for treatment response studies). "Moderate quality" studies were defined as studies having at least moderate quality ratings for all three items (with „low" or „high" risk of bias for item 8). All studies that received a low-quality rating in one or several items (items 1-7) or had an unknown risk of bias for item 8 were considered "low quality" studies.

Criteria for the quality assessment of reporting oculomotor findings in ataxia

Study cohort related items

1. Patient selection

high = underlying disorder in all patients included was confirmed by genetic testing

moderate = no genetic testing available but either positive family history with a clear pattern of inheritance autosomal dominant [AD], autosomal recessive [AR], X-linked recessive [XR]) or established and specific diagnostic biomarkers used

low = clinically- or imaging-based diagnosis, no genetic testing, no positive family history

2. Control group selection

high = control group age-matched, independent from test group, clinical assessment (exclusion of abnormal findings) provided in detail; patients serve as their own controls in longitudinal treatment trials or treatment and placebo groups were age-matched in placebo-controlled treatment trials.

moderate = age-matched, reportedly healthy but no details reported how this was assessed

low = one or several of the following: controls are not age-matched; no information about their age is available; no information about selection process is provided; controls are genetically undetermined relatives of the test patients included.

not available = no control group is provided

Items related to data acquisition

3. Rigidity of recording protocol applied

high = pre-specified recording protocol available for all parameters studied, identical protocol applied in all participants and sessions

moderate = pre-specified recording protocol available only for selected parameters or different pre-specified recording protocols applied in included patients and sessions.

low = no pre-specified recording protocol used or no information provided about the use of a recording protocol.

4. Description of recording parameters used

high = all recording parameters are reported with sufficient detail to reproduce the study

moderate = recording parameters are reported partially only

low = no information provided about recording parameters

5. Appropriateness of recording devices used for oculomotor testing

high = high temporal and spatial resolution, high signal-to-noise recording technique used (e.g. search coils, high-quality video-oculography with sampling rate of >=100Hz)

moderate = recording technique with moderate to high noise levels (e.g. electro-oculography, electro-nystagmography, low to middle performance video-oculography (sampling rate <100Hz)) or behavioral paradigm used (e.g. Subjective Visual Vertical)

low = no details about recording device reported or high noise levels described

Items related to data analysis

6. Description of data analysis

high = data analysis is described with sufficient detail to allow reproduction of study

moderate = data analysis description is provided, but details are lacking and thus is insufficient to allow reproduction of study

low = no information provided about how the data analysis was performed or inadequate analysis pipeline

7. Assessment of obtained test results for significance

high = statistical analysis using normative values obtained from a control group or baseline values (for longitudinal studies)

moderate = statistical analysis using normative values from published literature or from the manufacturer of the device used

low = no normative values available and/or no statistical analysis performed

8. Risk of bias for assessing test results

low = index test results interpreted in a blinded fashion (i.e., without knowledge if the underlying disorder was present or absent) or automated data analysis without rater's input.

high = index test results interpreted in a non-blinded fashion only and / or based on the rater's subjective assessment (e.g. rating of overall pattern of vHIT traces).

unclear = no information provided about blinding of reviewers of test results.

Appendix 3 – additional tables

Supplementary Table 1 – Meta-data of included studies listed in alphabetical order

Table S1: meta-data of selected studies

Author, year (citation)	Study population	Study location	Data collection (analysis)	Subjects (% females)	Mean age (SD)	Paradigms recorded	Recording device used	Clinical scale(s)	Special comments
Alexandre et al. 2013 [2]	FRDA, SCA1, SCA3	monocentric	Prospective, case-control-study	21 (43%)	48 (14)	VGS, SI,	VOG (mobile EBT)	SARA	
Anastasopoulos et al. 1998 [3]	mixed hereditary (SCA1, ADCA others) and non-hereditary	monocentric	Prospective, case-control-study	10 (NR)	39 (10)	PEM, OVAR,	Search coils	N/A	
Anderson et al. 2002a [4]	SCA2, EA2	monocentric	Prospective, case-series	4 (50%)	54 (13)	PEM, VGS,	Search coils	N/A	
Anderson et al. 2002b [5]	SCA8	monocentric	Prospective, case-control-study	3 (NR)	NR (NR)	rVOR, vVOR Tc	EOG	N/A	
Arpa et al. 1995 [6]	MSA-C, LOCA	monocentric	Prospective, case-series	40 (55%)	59 (10)	PEM, VGS, OKN, rVOR, VORs, SN, GEN, RBN,	EOG	N/A	
Baloh et al. 1975 [7]	FRDA, CA	monocentric	Retrospective, case-control-study	15 (NR)	NR (NR)	PEM, VGS, SI, OKN, rVOR, SN, PN, GEN, RBN, CI	EOG	N/A	
Baloh et al. 1978 [8]	A-T	monocentric	Prospective, case-series	6 (50%)	11 (7)	PEM, VGS, OKN, rVOR, VORs, SN, CI	EOG	N/A	
Bargagli et al. 2021 [9]	AOA2	Monocentric	Prospective, case-control-study	2 (50%)	11 (1)	VGS, AS, GEN, RBN	VOG (ASL 504 eye tracker)	N/A	

Bour et al. 2008 [10]	SCA6, ADCA others	monocentric	Prospective, case-control-study	11 (45%)	42 (15)	PEM, VGS, SI, SN, HN, GEN,	Search coils	N/A	
Bremova et al. 2016 [11]	NPC	monocentric	Prospective, case-control-study	8 (NR)	27 (10)	qHIT, oVEMPs, cVEMPs, CI, SVV	VOG (EyeSeeCam, Interacoustics)	SARA	
Bremova et al. 2015 [12]	NPC	monocentric	Prospective, non-randomized treatment study	12 (42%)	23 (5)	PEM, VGS, OKN, rVOR,	VOG (EyeSeeCam, Interacoustics)	SARA, SCAFI	
Brokalaki et al. 2015 [13]	Autoimmune (anti-GAD-ab positive) cerebellar ataxia	monocentric	Retrospective, single case report	1 (100%)	64	SI, GEN	VOR (IRIS)	SARA	
Bronstein et al. 2008 [14]	Midline cerebellar degeneration	monocentric	Prospective, case-control-study	8 (50%)	49 (NR)	rVOR Tc, self-motion perception decay Tc	EOG	N/A	
Brown et al. 1993 [15]	LOCA	monocentric	Prospective, case-control-study	5 (0%)	47 (10)	VGS,	EOG	N/A	
Bürk et al. 1996 [16]	SCA1, SCA2, SCA3	monocentric	Prospective, case-control-study	24 (NR)	SCA1=34 (4) SCA2=35 (5) SCA3=41 (4)	VGS	EOG	N/A	
Bürk et al. 1997 [17]	SCA1, SCA2, SCA3, ADCA others, LOCA	monocentric	Prospective, case-control-study	48 (NR)	44-58 (6-16)	VGS	EOG	N/A	
Büttner et al. 1998 [18]	SCA1, SCA2, SCA3, SCA6	monocentric	Prospective, case-series	20 (NR)	46 (12)	PEM, VGS, SI, OKN, rVOR, VVOR, VORs, SN, GEN, RBN,	EOG	N/A	
Caspi et al. 2013 [19]	SCA3	monocentric	Prospective, case-control-study	10 (70%)	49 (15)	VGS	Search coils	SARA	

Ceravolo et al. 2002 [20]	LOCA	monocentric	Prospective, case-control-study	36 (NR)	LOCA=47 (14) LOCA+=50 (20)	PEM, VGS, SI, PN, GEN, RBN, CI,	EOG	ICARS	
Chang et al. 2020 [21]	FRDA, SCA1, SCA2, SCA3, SCA6, other SCA (not specified), AT, HSP, ARCA, MSA-C	monocentric	Prospective, case-control-study	59 (54%)	53 (19)	PEM	VOG (smartphone)	BARS	
Choi et al. 2015 [22]	EA2	monocentric	Prospective, case-series	4 (50%)	26 (12)	SN	VOG (SMI)	N/A	
Christova et al. 2008 [23]	SCA6	monocentric	Prospective, case-control-study	9 (55%)	43 (12)	PEM, VGS, SI,	Search coils	ICARS	
Ciuffreda et al. 1985 [24]	FRDA	monocentric	Retrospective, single case report	1 (100%)	23	PEM, VGS, SI	EOG	N/A	
Clausi et al. 2013 [25]	AOA2	monocentric	Prospective, case-control-study	2 (50%)	39 (1)	VGS, SI,	VOG (NR)	ICARS	
Coin and Vance 2021 [26]	EA4	monocentric	Prospective, non-randomized treatment study	3 (67%)	52 (14)	PEM, GEN	VOG (consumer digital camera)	N/A	
Crane et al. 2000 [27]	FRDA, SCA3, SCA6, ARCA, EOCA	monocentric	Prospective, case-control-study	11 (NR)	48 (16)	rVOR, tVOR	Search coils	N/A	
Crowdy et al. 2000 [28]	SCA1, SCA7, ADCA others, EOCA, LOCA	monocentric	Prospective, case-control-study	8 (25%)	43 (9)	VGS	VOG (NR)	N/A	
Dakin et al. 2018 [29]	SCA6	monocentric	Prospective, case-control-study	15 (47%)	67 (10)	SVV	NA	SARA	
Dale et al. 1978 [30]	FRDA	Monocentric	Retrospective, case series	2 (50%)	19 (2)	PEM, SI, OKN, GEN,	EOG	N/A	

de Oliveira et al. 2021 [31]	SCA3 (symptomatic/pre-symptomatic)	monocentric	Prospective, case-control-study	73 (52%)	Symp=42 (9) presymp <4y= 33 (9) Presymp >4y=27 (6)	PEM, VGS, SI, GEN, qHIT	VOG (eyeseecam, Interacoustics)	CCFS, ICARS, INAScount, NESSCA, SARA,	38 pre-symptomatic carriers included (est. onset <4 years or >4 years)
Ell et al. 1984 [32]	FRDA	Monocentric	Prospective, case series	10 (40%)	25 (9)	PEM, VGS, SI, OKN, VORs, CI,	EOG	N/A	
Fahey et al. 2008 [33]	FRDA	monocentric	Prospective, case-control-study	20 (NR)	35 (13)	PEM, VGS, SI, SN, GEN, qHIT	VOG (Micromedical Technologies) or search coils	FARS, VF14, VFQ39, SLCLC	
Federighi et al. 2011 [34]	SCA2, LOCA	monocentric	Prospective, case-control-study	15 (40%)	SCA2=44 LOCA=51	VGS, AS,	VOG (ASL 504 eye tracker)	ICARS	
Federighi et al. 2017 [35]	ATLD	monocentric	Prospective, case-control-study	2 (50%)	45 (1)	VGS, SI, GEN, RBN	VOG (ASL 504 eye tracker)	ICARS	
Fetter et al. 1994 [36]	LOCA	monocentric	Prospective, case-control-study	17 (NR)	LOCA=54 (6) LOCA+=58 (9)	PEM, VGS, SI, OKN, rVOR, rVOR decay Tc, VORs, SN, GEN, RBN,	EOG	N/A	
Fielding et al. 2010 [37]	FRDA	monocentric	Prospective, case-control-study	13 (NR)	36 (9)	VGS, MGS, AS	Search coils	FARS, SLCLC	
Furman et al. 1986 [38]	EOCA	Monocentric	Prospective, case series	4 (NR)	13 (13)	PEM, VGS, SI, rVOR, VVOR, VORs, SN	EOG or search coils	N/A	
Furman et al. 1983 [39]	FRDA	monocentric	Prospective, case-control-study	24 (54%)	NR	PEM, VGS, SI, OKN, rVOR, VVOR, VORs, GEN, RBN	EOG	N/A	

Geisinger et al. 2021 [40]	SCA3	Monocentric	Prospective, case-control-study	21 (71%)	59 (16)	qHIT, cVEMPs	VOG (ICS impulse goggles, Natus)	SARA	
Ghasia et al. 2016 [41]	SCA3	Monocentric	Prospective, case series	12 (67%)	50 (17)	PEM, VGS, SI, GEN	VOR (eyelink 1000 or Jazz Novo)	SARA	
Gomez et al. 1997 [42]	SCA6	Monocentric	Prospective, case series	13 (NR)	NR	PEM, VGS, SI, rVOR, rVOR decay Tc, VORs, SN, GEN,	Search coils	N/A	
Gonzalez-Martin et al. 2004 [43]	EOCA	Monocentric	Prospective, case series	2 (100%)	11 (0)	Reflexive saccades triggered by head rotations	EOG	N/A	
Gordon et al. 2014 [44]	SCA3	monocentric	Prospective, case-control-study	10 (70%)	49 (15)	qHIT	Search coils	SARA	Same patients as in Caspi et al. 2013 [19]
Costales et al. 2021 [45]	RFC1-related ataxia	Monocentric	Retrospective, case series	11 (64%)	69 (4)	SN, qHIT	VOG (ICS impulse goggles, Natus)	SARA	
Havla et al. 2020 [46]	NPC	monocentric	Prospective, case-control-study	31 (48%)	NPC1-P=24 NPC1-MC=50	PEM, VGS,	VOG (eyeseecam, Interacoustics)	SARA, SCAFI, SLCLC	Including 17 asymptomatic mutation carriers
Helmchen et al. 2017 [47]	Other signs of cerebellar disease (DBN)	monocentric	Prospective, case-control-study	27 (NR)	74 (9)	PEM, SN	VOG (eyelink II, SR Research	SARA	
Hocking et al. 2014 [48]	FRDA	monocentric	Prospective, case-control-study	13 (NR)	36 (9)	VGS	Search coils	N/A	Same patients as in Fielding et al. 2010 [37]
Hocking et al. 2010 [49]	FRDA	monocentric	Prospective, case-control-study	13 (NR)	36 (9)	VGS	Search coils	FARS, SLCLC	Same patients as in Fielding et al. 2010 [37]

Hübner et al. 2007 [50]	SCA17	monocentric	Prospective, case-control-study	15 (27%)	37 (11)	PEM, VGS, MGS, SN, GEN, RBN,	VOG (eyelink II, SR Research	N/A	
Huh et al. 2015 [51]	SCA6	monocentric	Prospective, case-control-study	11 (45%)	59 (12)	PEM, rVOR, qHIT, CI	EOG or search coils	ICARS	
Joiner et al. 2005 [52]	SCA6	Monocentric	Prospective, case series	3 (NR)	NR	VGS	VOG (NR)	N/A	
Jorge et al. 2020 [53]	Acute-onset cerebellar ataxia	Monocentric	Prospective, single case report	1 (100%)	37	PEM, SN, PN, GEN, qHIT	VOG (VO425, Interacoustics)	N/A	
Kalla et al. 2011 [54]	Other signs of cerebellar disease (DBN)	monocentric	Prospective, non-randomized treatment study	8 (75%)	68 (6)	SN, GEN	VOG (GN Otometrics Hortmann Vestlab 100)	N/A	
Kattah et al. 1983 [55]	CA	Monocentric	Prospective, case series	3 (100%)	29 (14)	PEM, VGS, OKN, rVOR, VORs, SN, PN, GEN,	EOG	N/A	
Kerber et al. 2005 [56]	SCA1, SCA6, SCA8, LOCA	Monocentric	Prospective, case series	20 (NR)	NR	PEM, VGS, OKN, rVOR, VORs	EOG	N/A	
Kim et al. 2013 [57]	FRDA, SCA1, SCA2, SCA3, SCA6, SCA7, SCA8, ADCA others	monocentric	Prospective, case-control-study	48 (44%)	49-57 (5-13)	PEM, VGS, SI, SN, HSN, PN, GEN,	VOR (Micromedical Technologies)	ICARS	
King et al. 2011 [58]	SCASI	monocentric	Prospective, case-control-study	2 (NR)	61 (NR)	PEM, VGS,	Search coils	N/A	
Kremmyda et al. 2012 [59]	CA	Monocentric	Prospective, case series	16 (25%)	72 (7)	PEM, rVOR, VVOR, VORs,	Search coils	N/A	
Kumar et al. 2005 [60]	SCASI, late-onset Tay Sachs, OPT	monocentric	Prospective, case-control-study	6 (NR)	NR	VGS	Seach coils	N/A	

Lasker et al. 2005 [61]	SCA6, CA	monocentric	Prospective, case-control-study	13 (NR)	NR	VGS	Search coils	N/A	
Lee et al. 2018 [62]	Wernicke Encephalopathy	Monocentric	Retrospective, case series	5 (20%)	58 (15)	qHIT	VOG (ICS impulse goggles, Natus)	N/A	
Lemos et al. 2018 [63]	SCA3	Monocentric	Prospective, single case report	1 (0%)	45	VGS, SI, GEN,	VOG (VO425, Interacoustics)	N/A	
Lewis and Crawford 2002 [64]	A-T	Monocentric	Prospective, case series	3 (NR)	NR	PEM, VGS,	Search coils	N/A	
Lewis et al. 1999 [65]	A-T	Monocentric	Prospective, case series	33 (NR)	10 (NR)	PEM, VGS, SI, OKN, rVOR,	EOG	A-T index score	
Lopez et al. 2019 [66]	FRDA, CA	Monocentric	Prospective, case series	4 (25%)	57 (8)	VGS	EOG	N/A	
Luis et al. 2016 [67]	FRDA, SCA1, SCA2, SCA3	Monocentric	Prospective, case-control-study	30 (50%)	FRDA=36 (12) SCA1=49 (17) SCA2=47 (15) SCA3=50 (12)	qHIT	VOG (eyeseecam, Interacoustics)	SARA	
Mariani et al. 2017 [68]	AOA1, AOA2, AT	Monocentric	Prospective, case-control-study	40 (45%)	AOA1=37 (NR) AOA2=38 (NR) AT=33 (NR)	PEM, VGS, AS, SI, SN, GEN	VOG (SMI)	SARA	
Matsuda et al. 2014 [69]	SCA6, SCA31	monocentric	Prospective, case-control-study	18 (56%)	64 (12)	SEM (visual search task)	VOG (Eyelink II, SR research)	N/A	
Matsuda et al. 2015 [70]	SCA6, SCA31	monocentric	Prospective, case-control-study	19 (53%)	65 (12)	SEM (visual search task)	VOG (Eyelink II, SR research)	N/A	Overlap with Matsuda et al. 2014 [69]
Migliaccio et al. 2004 [71]	CA	Monocentric	Prospective, case series	4 (NR)	NR	PEM, VGS, OKN, rVOR, VVOR, VORs GEN, qHIT	Search coils	N/A	

Migliaccio and Watson 2016 [72]	CANVAS	Monocentric	Prospective, single case report	1 (100%)	68	PEM, VGS, VORs, qHIT	Search coils	N/A	
Moreno-Ajona et al. 2019 [73]	CANVAS	Monocentric	Prospective, case series	5 (60%)	70 (12)	VVOR, VORs, qHIT, CI, oVEMPs, cVEMPs	VOG (ICS impulse goggles, Natus)	N/A	
Morrow et al. 1992 [74]	CA	monocentric	Prospective, case-control-study	19 (NR)	38 (NR)	PEM, OKN, VORs	EOG	N/A	
Moschner et al. 1994 [75]	FRDA, OPCA, CA	monocentric	Prospective, case-control-study	71 (45%)	FRDA=21 (10) OPCA=47 (14) CA=43 (15)	PEM, VGS, SI, OKN, rVOR, VORs, SN, GEN	EOG	N/A	
Oh et al. 2001 [76]	SCA7	Monocentric	Prospective, case series	2 (100%)	50 (23)	PEM, VGS, rVOR, VORs, GEN	EOG	N/A	
Ohyagi et al. 2000 [77]	SCA3	monocentric	Prospective, case-control-study	8 (63%)	36 (14)	NA	NA	N/A	Synoptophere used to assess vergence
Pretegiani et al. 2018 [78]	SCA2, LOCA	monocentric	Prospective, case-control-study	22 (36%)	SCA2=38.7 LOCA=42.4	AS	VOG (ASL 504 eye tracker)	ICARS	
Reetz et al. 2018 [79]	SCA2	monocentric	Prospective, case-control-study	26 (38%)	42 (10)	VGS	EOG	SARA	
Rey-Martinez et al. 2018 [80]	CANVAS	monocentric	Prospective, case-control-study	5 (40%)	70 (NR)	VVOR	VOG (ICS impulse goggles, Natus)	N/A	
Ribai et al. 2007 [81]	FRDA	monocentric	Prospective, non-randomized treatment study	37 (NR)	NR	SI	EOG	ICARS	
Ribeiro et al. 2015 [82]	SCA3	monocentric	Prospective, case-control-study	14 (57%)	42 (8)	oVEMPs, cVEMPs	NA	N/A	

Rodríguez-Díaz et al. 2018 [83]	SCA2	monocentric	Prospective, randomized treatment study	38 (39%)	39 (11)	VGS	EOG	SARA	
Rodríguez-Labrada et al. 2017 [84]	SCA2	monocentric	Prospective, case-control-study	48 (40%)	41 (10)	VGS	EOG	SARA	
Rodríguez-Labrada et al. 2016 [85]	SCA2	monocentric	Prospective, observational study	30 (27%)	40 (11)	VGS	EOG	SARA	
Rosini et al. 2013 [86]	ADCA others	monocentric	Prospective, non-randomized treatment study	2 (100%)	59 (1)	VGS, SI	VOG (ASL 504 eye tracker)	ICARS	
Rufa and Federighi 2011 [87]	SCA2, LOCA	Monocentric	Prospective, case series	18 (39%)	SCA2=47 LOCA=51	VGS	VOG (ASL 504 eye tracker)	N/A	
Sağlam and Lehnen 2014 [88]	SCA2, LOCA	monocentric	Prospective, case-control-study	9 (67%)	57 (13)	qHIT	Search coils	SARA	
Samuel et al. 2004 [89]	CA	Monocentric	Prospective, case series	6 (0%)	58 (10)	PEM, VGS, rVOR, VVOR, SN	EOG	N/A	
Seifried et al. 2005 [90]	SCA2	Monocentric	Prospective, observational study	82 (37%)	NR	VGS	EOG	N/A	
Serrano-Munuera et al. 2013 [91]	SCA37	monocentric	Prospective, case-control-study	2 (NR)	67 (4)	PEM, VGS, OKN	EOG	SARA	
Shaikh et al. 2013 [92]	A-T	Multicentric	Prospective, case-control-study	13 (46%)	26 (NR)	rVOR, rVOR decay Tc, cVEMPs	VOG (SMI) or search coils	N/A	
Shaikh et al. 2011 [93]	A-T	Multicentric	Prospective, case series	13 (46%)	26 (NR)	SI, SN, GEN	VOG (SMI) or search coils	N/A	
Shaikh et al. 2009 [94]	A-T	Multicentric	Prospective, non-randomized treatment study	4 (NR)	35 (14)	rVOR, rVOR decay Tc, SN	Search coils	N/A	

Solomon et al. 2005 [95]	NPC	Monocentric	Prospective, single case report	1 (NR)	NR	VGS	Search coils	N/A	
Spieker et al. 1995 [96]	FRDA	Monocentric	Prospective, case-control-study	13 (38%)	30 (7)	PEM, VGS, SI, OKN, rVOR, rVOR decay Tc, VORs, SN, GEN, RBN, CI	EOG	N/A	
Szmulewicz et al. 2011 [97]	CANVAS	Monocentric	Prospective, case series	27 (47%)	71 (NR)	CI, qHIT	VOG (ICS impulse goggles, Natus)	N/A	
Takegoshi and Murofushi 2000 [98]	SCA3, CA	Monocentric	Prospective, case-control-study	16 (63%)	57 (11)	CI, cVEMPs	EOG	N/A	
Takeichi et al. 2000 [99]	SCA6	Monocentric	Prospective, case-control-study	5 (20%)	57 (8)	PEM, rVOR, VVOR, VORs,	VOG (unclear)	N/A	
Tarnutzer et al. 2016 [100]	CANVAS	Monocentric	Prospective, case series	5 (40%)	72 (9)	qHIT	VOG (ICS impulse goggles, Natus)	N/A	
Terao et al. 2016 [101]	SCA6, SCA8, SCA31, MSA-C, CA	Monocentric	Prospective, case-control-study	41 (56%)	SCA=66 (10) MSA-C=63 (8)	VGS, MGS	EOG	N/A	
Terao et al. 2017 [102]	SCA6, SCA8, SCA31, MSA-C	Monocentric	Prospective, case-control-study	44 (NR)	SCD=65 (10) MSA-C=63 (7)	VGS, MGS	EOG	N/A	
Tilikete et al. 2005 [103]	Autoimmune (anti-GAD-ab positive) cerebellar ataxia	monocentric	Retrospective, single case report	1 (100%)	76	rVOR, SN	VOG (Ulmer, Synapsys)	ICARS	
Velázquez-Pérez et al. 2011 [104]	SCA2	Monocentric	Prospective, randomized treatment study	33 (NR)	42 (6)	VGS	EOG	SARA	

Velázquez-Pérez et al. 2012 [105]	SCA2	Monocentric	Prospective, non-randomized treatment study	12 (25%)	38 (9)	VGS	EOG	SARA	
Velázquez-Pérez et al. 2014 [106]	SCA2	Monocentric	Prospective, case-control-study	37 (65%)	40 (12)	AS	EOG	SARA	
Velázquez-Pérez et al. 2009 [107]	SCA2	Monocentric	Prospective, case-control-study	54 (63%)	36 (NR)	VGS	EOG	SARA	
Versino et al. 2009 [108]	CA with opsoclonus	monocentric	Retrospective, single case report	1 (0%)	26	VGS	VOG (IRIS, Scalar)	N/A	
Walterfang et al. 2013 [109]	NPC	Monocentric	Prospective, case-control-study	10 (40%)	32 (10)	VGS	VOG (Microguide 1000 infrared limbus)	N/A	Same patients as in Walterfang et al. 2013 [109]
Walterfang et al. 2012 [110]	NPC	Monocentric	NA, case-control-study	10 (40%)	32 (10)	VGS, AS	VOG (Microguide 1000 infrared limbus)	N/A	
Wessel et al. 1998 [111]	FRDA, SCA1, SCA3, CA, OPCA	Monocentric	Prospective, case-control-study	26 (NR)	FRDA=39 (NR) CA=52 (NR) OPCA=43 (NR)	PEM, VGS, SI, OKN, rVOR, rVOR decay Tc, VORs, SN, REN, RBN,	EOG	N/A	
Wiest et al. 2001 [112]	SCA6, EA2	Monocentric	Prospective, case-control-study	6 (NR)	59 (13)	tVOR	Search coils	N/A	
Wu et al. 2017 [113]	SCA3 (symptomatic/pre-symptomatic)	Monocentric	Prospective, case-control-study	Pre-SCA3=6 (50%) SCA3=23 (52%)	Pre-SCA3= 29.8±7.4 SCA3= 39.8±10.9	PEM, VGS, AS, SI, GEN,	VOG (VO425, Interacoustics)	SARA	
Yacovino et al. 2019 [114]	CANVAS	Monocentric	Retrospective, case-control-study	5 (60%)	73 (7)	VVOR, VORs, CI, qHIT,	VOG (eyeseecam, Interacoustics)	N/A	

						oVEMPs, cVEMPs			
Yee et al. 1992 [115]	LOCA (Gerstmann-Sträussler-Scheinker Disease)	Monocentric	Prospective, case-control-study	5 (80%)	61 (10)	PEM, VGS, OKN, rVOR, VVOR, VORs, GEN, RBN,	EOG or search coils	N/A	
Yu et al. 1990 [116]	CA	Monocentric	Prospective, case-control-study	8 (13%)	30 (NR)	PEM	EOG or search coils	N/A	
Yue et al. 1997 [117]	ADCA others	Monocentric	Prospective, case series	4 (NR)	NR	PEM, VGS, OKN, rVOR, VORs,	EOG	N/A	
Zee et al. 1976 [118]	ADCA others	Monocentric	Prospective, case series	12 (63%)	64 (7)	PEM, VGS, OKN, rVOR, VVOR, VORs, SN, GEN, RBN,	EOG or VOG (custom made)	N/A	

Abbreviations: ADCA=autosomal-dominant cerebellar ataxia; ARCA=autosomal-recessive cerebellar ataxia; AS=anti-saccades; A-T=ataxia telangiectasia; ATLD=ataxia telangiectasia like disease; BARS=brief ataxia rating scale; CA=cerebellar ataxia (not further specified); CCFS=Composite Cerebellar Functional Score; CI=caloric irrigation; cVEMPs=cervical vestibular-evoked myogenic potentials; EA=episodic ataxia; EOCA=early-onset cerebellar ataxia; EOG=electro-oculography; FRDA=Friedreich Ataxia; FARS=Friedreich's Ataxia Rating Scale; GEN=gaze-evoked nystagmus; HN=hyperventilation nystagmus; HSN=head-shaking nystagmus; ICARS=International Cooperative Ataxia Rating Scale; INAScount=Inventory of Non-Ataxia Signs; LOCA=late-onset cerebellar ataxia; MGS=memory-guided saccades; NESSCA= Neurological Examination Score for Spinocerebellar Ataxia; NPC=Niemann Pick disease Type C; OKN=optokinetic nystagmus; OPCA=olivoponto cerebellar ataxia; OPT=oculopalatal tremor; OVAR=off-vertical axis rotation; oVEMPs=ocular vestibular-evoked myogenic potentials; PEM=pursuit eye movements; PN=positional nystagmus; qHIT=quantitative head-impulse test; RBN=rebound nystagmus; rVOR=rotational vestibulo-ocular reflex; SARA=Scale for the Assessment and Rating of Ataxia; SCA=spinocerebellar ataxia; SCAFI=Spinocerebellar ataxia functional index; SCASI=spinocerebellar ataxia with saccadic intrusions; SEM=saccadic eye movements; SI=saccadic intrusions; SLCLC=Sloan Low-Contrast Letter Chart; SN=spontaneous nystagmus; SVV=subjective visual vertical; tVOR=translational vestibulo-ocular reflex; VF14 and VFQ39=visual quality of life scales; VOG=video-oculography; VORs=vestibulo-ocular reflex suppression; VGS=visually-guided saccades; VVOR=visually-enhanced vestibulo-ocular reflex;

Table S2: quality assessment for studies reporting on oculomotor findings in ataxia – overview on all selected studies

Table S2: quality assessment for studies reporting on oculomotor findings in ataxia – overview on all selected studies									
Study	**Patient selection**	**Control group selection**	**Rigidity of recording protocol applied**	**Description of recording parameters used**	**Appropriateness of recording devices used for OM /vestibular testing**	**Appropriateness of data analysis applied**	**Assessment of obtained test results for significance**	**Risk of bias for assessing test results**	**OVERALL study quality rating**
Alexandre et al. 2013 [2]	high	moderate	high	high	high	moderate	high	low	moderate
Anastasopoulos et al. 1998 [3]	low	moderate	high	high	high	high	high	high	low
Anderson et al. 2002a [4]	moderate	moderate	high	high	high	moderate	high	low	moderate
Anderson et al. 2002b [5]	high	low	high	high	moderate	high	high	low	low
Arpa et al. 1995 [6]	low	low	high	high	moderate	moderate	low	high	low
Baloh et al. 1975 [7]	low	low	high	low	moderate	moderate	low	high	low
Baloh et al. 1978 [8]	low	low	high	low	moderate	low	low	unclear	low
Bargagli et al. 2021 [9]	high	moderate	high	high	high	high	high	low	high
Bour et al. 2008 [10]	high	moderate	high	high	high	high	high	low	high
Bremova et al. 2016 [11]	moderate	moderate	high	high	high	high	high	low	moderate
Bremova et al. 2015 [12]	moderate	high	high	moderate	high	low	high	low	moderate
Brokalaki et al. 2015 [13]	low	high	high	moderate	high	low	low	high	low
Bronstein et al. 2008 [14]	low	moderate	high	high	moderate	high	high	low	low
Brown et al. 1993 [15]	low	moderate	high	high	moderate	high	high	low	low
Bürk et al. 1996 [16]	high	moderate	high	moderate	moderate	moderate	high	unclear	low
Bürk et al. 1997 [17]	moderate	moderate	high	moderate	moderate	moderate	high	unclear	low
Büttner et al. 1998 [18]	high	not applicable	high	high	moderate	high	low	high	moderate

Caspi et al. 2013 [19]	high	moderate	high	high	high	high	high	unclear	low
Ceravolo et al. 2002 [20]	low	low	high	moderate	moderate	moderate	moderate	low	low
Chang et al. 2020 [21]	low	low	high	high	moderate	high	high	low	low
Choi et al. 2015 [22]	high	not applicable	high	moderate	moderate	low	low	unclear	low
Christova et al. 2008 [23]	high	moderate	high	high	high	moderate	high	low	moderate
Ciuffreda et al. 1985 [24]	low	not applicable	low	moderate	moderate	low	low	high	low
Clausi et al. 2013 [25]	high	moderate	high	high	high	moderate	high	unclear	low
Coin and Vance 2021 [26]	high	high	low	low	moderate	low	low	high	low
Costales et al. 2021 [45]	high	not applicable	high	moderate	high	moderate	low	low	low
Crane et al. 2000 [27]	low	low	high	high	high	high	high	low	low
Crowdy et al. 2000 [28]	low	high	high	high	high	high	high	low	low
Dakin et al. 2018 [29]	high	moderate	high	high		high	high	low	high
Dale et al. 1978 [30]	moderate	not applicable	high	moderate	moderate	moderate	low	high	moderate
de Oliveira et al. 2021 [31]	high	low	high	moderate	high	high	high	low	low
Ell et al. 1984 [32]	moderate	not applicable	high	moderate	moderate	moderate	low	high	moderate
Fahey et al. 2008 [33]	high	low	high	high	high	high	high	low	low
Federighi et al. 2011 [34]	high	high	high	high	high	high	high	low	high
Federighi et al. 2017 [35]	high	moderate	high	high	high	high	high	low	high
Fetter et al. 1994 [36]	low	moderate	high	high	moderate	high	high	low	low
Fielding et al. 2010 [37]	high	moderate	high	high	high	high	high	low	high
Furman et al. 1986 [38]	moderate	not applicable	high	high	high	low	low	high	low
Furman et al. 1983 [39]	low	low	high	high	moderate	low	high	unclear	low
Geisinger et al. 2021 [40]	high	high	high	high	high	high	high	low	high

Ghasia et al. 2016 [41]	high	low	high	moderate	high	high	moderate	high	low
Gomez et al. 1997 [42]	high	not applicable	high	high	high	moderate	low	high	low
Gonzalez-Martin et al. 2004 [43]	low	not applicable	low	low	moderate	low	low	high	low
Gordon et al. 2014 [44]	high	moderate	high	high	high	high	high	low	high
Havla et al. 2020 [46]	high	moderate	high	high	high	high	high	low	high
Helmchen et al. 2017 [47]	low	moderate	high	high	high	low	high	low	low
Hocking et al. 2014 [48]	high	moderate	high	high	high	high	high	low	high
Hocking et al. 2010 [49]	high	moderate	high	high	high	high	high	low	high
Hübner et al. 2007 [50]	high	moderate	high	moderate	high	high	high	low	moderate
Huh et al. 2015 [51]	high	moderate	high	high	high	high	high	low	high
Joiner et al. 2005 [52]	high	low	high	high	low	moderate	low	low	low
Jorge et al. 2020 [53]	low	high	high	high	high	moderate	low	low	low
Kalla et al. 2011 [54]	low	high	high	high	moderate	moderate	high	low	moderate
Kattah et al. 1983 [55]	moderate	low	high	moderate	moderate	low	low	unclear	low
Kerber et al. 2005 [56]	high	not applicable	high	moderate	moderate	moderate	moderate	high	moderate
Kim et al. 2013 [57]	high	moderate	high	high	high	high	high	high	moderate
King et al. 2011 [58]	low	low	high	high	high	moderate	high	low	low
Kremmyda et al. 2012 [59]	low	not applicable	high	high	high	high	high	low	low
Kumar et al. 2005 [60]	low	moderate	high	high	high	high	high	low	low
Lasker et al. 2005 [61]	moderate	moderate	high	high	high	moderate	high	low	moderate
Lee et al. 2018 [62]	low	not applicable	high	high	high	high	moderate	low	low
Lemos et al. 2018 [63]	high	not applicable	high	high	high	low	low	low	low

Lewis and Crawford 2002 [64]	moderate	not applicable	high	moderate	high	high	low	low	low
Lewis et al. 1999 [65]	moderate	low	high	moderate	moderate	high	moderate	low	low
Lopez et al. 2019 [66]	low	moderate	low	moderate	moderate	moderate	high	low	low
Luis et al. 2016 [67]	high	high	high	high	high	high	high	low	high
Mariani et al. 2017 [68]	high	moderate	high	high	high	high	high	low	high
Matsuda et al. 2014 [69]	moderate	moderate	high	high	high	high	high	low	high
Matsuda et al. 2015 [70]	high	moderate	high	moderate	high	high	high	low	moderate
Migliaccio et al. 2004 [71]	low	low	high	high	high	high	low	low	low
Migliaccio and Watson 2016 [72]	low	low	high	high	high	high	low	low	low
Moreno-Ajona et al. 2019 [73]	low	low	high	moderate	high	moderate	moderate	low	low
Morrow et al. 1992 [74]	low	moderate	high	high	moderate	moderate	high	low	low
Moschner et al. 1994 [75]	low	moderate	high	high	moderate	moderate	high	low	low
Oh et al. 2001 [76]	high	low	high	high	moderate	moderate	moderate	low	low
Ohyagi et al. 2000 [77]	high	low	high	moderate		high	low	low	low
Pretegiani et al. 2018 [78]	high	moderate	high	high	high	high	high	low	high
Reetz et al. 2018 [79]	high	moderate	high	high	moderate	high	high	low	moderate
Rey-Martinez et al. 2018 [80]	low	low	high	high	high	high	high	low	low
Ribai et al. 2007 [81]	high	high	high	high	moderate	moderate	high	unclear	low
Ribeiro et al. 2015 [82]	high	moderate	high	high		high	high	low	high
Rodríguez-Díaz et al. 2018 [83]	high	high	high	high	moderate	high	high	low	moderate

Rodríguez-Labrada et al. 2017 [84]	high	moderate	high	high	moderate	moderate	high	low	moderate
Rodríguez-Labrada et al. 2016 [85]	high	high	high	high	moderate	high	high	low	moderate
Rosini et al. 2013 [86]	low	high	high	high	high	high	high	low	high
Rufa and Federighi 2011 [87]	high	moderate	high	high	high	high	high	low	high
Sağlam and Lehnen 2014 [88]	low	low	high	high	high	high	high	low	low
Samuel et al. 2004 [89]	low	not applicable	high	high	high	moderate	low	high	low
Seifried et al. 2005 [90]	high	moderate	high	high	moderate	high	high	low	moderate
Serrano-Munuera et al. 2013 [91]	high	low	high	high		high	moderate	low	low
Shaikh et al. 2013 [92]	moderate	low	moderate	high	moderate	high	high	low	low
Shaikh et al. 2011 [93]	moderate	not applicable	moderate	low	moderate	moderate	low	unclear	low
Shaikh et al. 2009 [94]	high	low	high	high	high	high	high	low	low
Solomon et al. 2005 [95]	moderate	not applicable	high	moderate	high	low	low	unclear	low
Spieker et al. 1995 [96]	moderate	high	high	high	moderate	high	high	low	moderate
Szmulewicz et al. 2011 [97]	low	low	high	high	high	moderate	low	high	low
Takegoshi and Murofushi 2000 [98]	high	low	high	high	moderate	high	high	low	low
Takeichi et al. 2000 [99]	high	moderate	high	high		high	high	low	high
Tarnutzer et al. 2016 [100]	low	not applicable	high	moderate	high	high	high	low	low
Terao et al. 2016 [101]	low	moderate	high	high	moderate	high	high	low	low
Terao et al. 2017 [102]	low	moderate	high	high	moderate	high	high	low	low
Tilikete et al. 2005 [103]	low	not applicable	high	moderate	moderate	low	low	low	low

Velázquez-Pérez et al. 2011 [104]	high	high	high	high	moderate	high	high	low	moderate
Velázquez-Pérez et al. 2012 [105]	high	high	high	high	moderate	high	high	low	moderate
Velázquez-Pérez et al. 2014 [106]	high	moderate	high	high	moderate	high	high	low	moderate
Velázquez-Pérez et al. 2009 [107]	high	high	high	high	moderate	high	high	low	moderate
Versino et al. 2009 [108]	low	high	high	moderate	moderate	low	low	unclear	low
Walterfang et al. 2013 [109]	moderate	moderate	high	high	high	high	high	low	moderate
Walterfang et al. 2012 [110]	moderate	moderate	high	high	high	high	high	low	moderate
Wessel et al. 1998 [111]	low	moderate	high	high	moderate	high	high	low	low
Wiest et al. 2001 [112]	high	moderate	high	high	high	high	high	low	high
Wu et al. 2017 [113]	high	high	high	high	high	high	high	high	high
Yacovino et al. 2019 [114]	low	low	high	moderate	high	moderate	low	low	low
Yee et al. 1992 [115]	moderate	low	high	moderate	moderate	moderate	low	low	low
Yu et al. 1990 [116]	low	moderate	high	high	moderate	high	high	low	low
Yue et al. 1997 [117]	high	low	high	high	moderate	low	low	low	low
Zee et al. 1976 [118]	moderate	low	high	high	high	low	low	high	low

Appendix 4 – additional figures

Figure S1: Normative values of smooth pursuit (SP) velocity gain in relation to target frequency. The mean and standard deviation values are indicated with circles and error bars, respectively, and are plotted against the target frequency. These data were extracted from to n=12 studies for horizontal eye movements and n=3 for vertical movements. The range of target frequencies that we recommend in our guidelines is highlighted as a gray area.

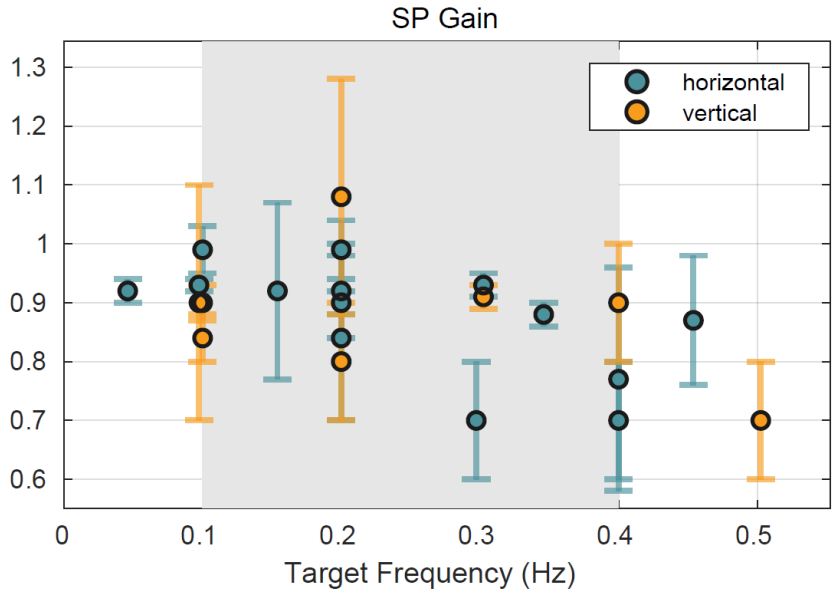


Figure S2: Normative values of horizontal saccadic peak velocity in relation to target jump amplitude. The mean and standard deviation values are indicated with circles and error bars, respectively, and are plotted against the target amplitude. The values were extracted from n=28 studies. The range of target jump amplitudes that we recommend in our guidelines is highlighted as a gray area.

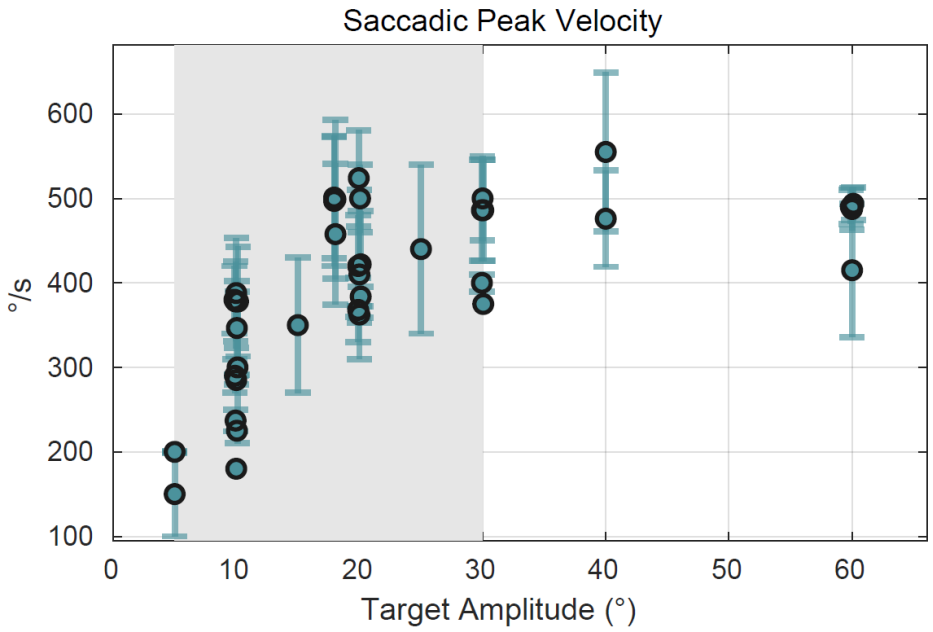


Figure S3: Normative values of horizontal saccade gain in relation to target jump amplitude. The mean and standard deviation values are indicated with circles and error bars, respectively, and are plotted against the target amplitude. The range of target jump amplitudes that we recommend in our guidelines is highlighted as a gray area.

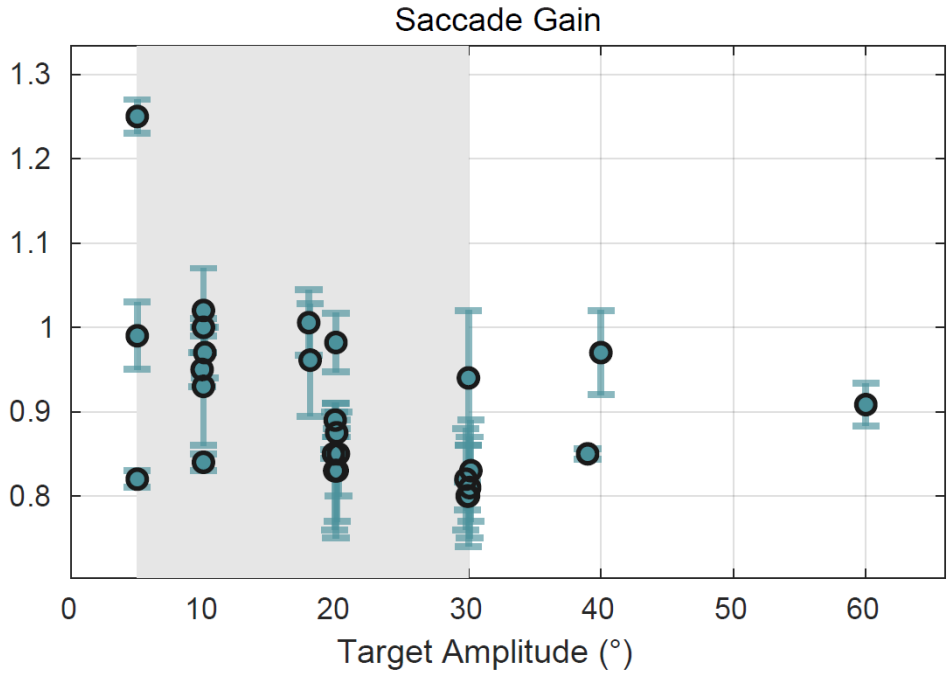


Figure S4: Normative values of horizontal saccadic latency in relation to target jump amplitude. The mean and standard deviation values are indicated with circles and error bars, respectively, and are plotted against the target amplitude. The values were extracted from n=15 studies. The range of target jump amplitudes that we recommend in our guidelines is highlighted as a gray area.

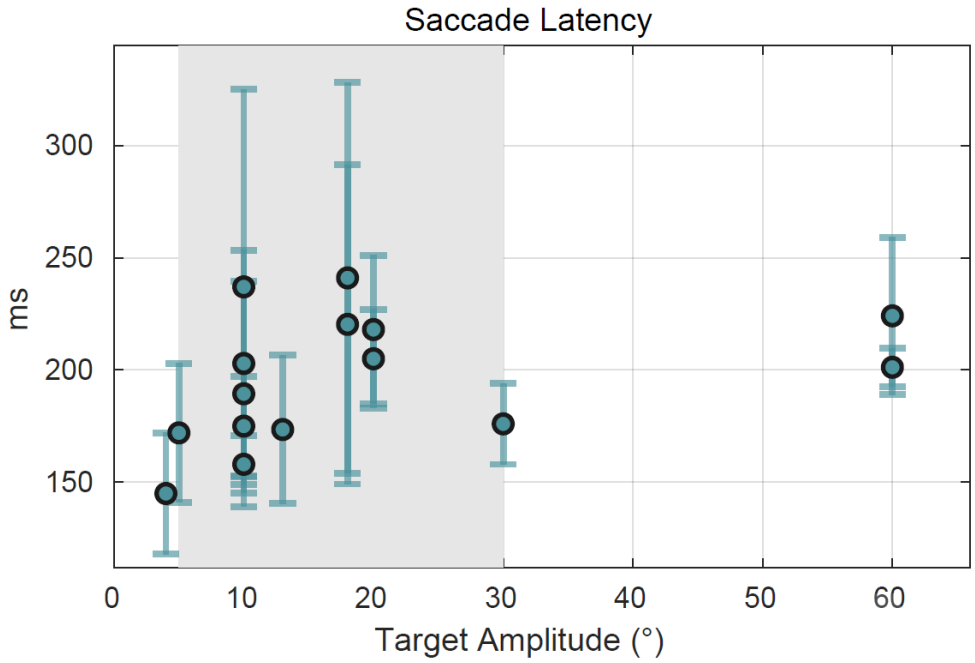
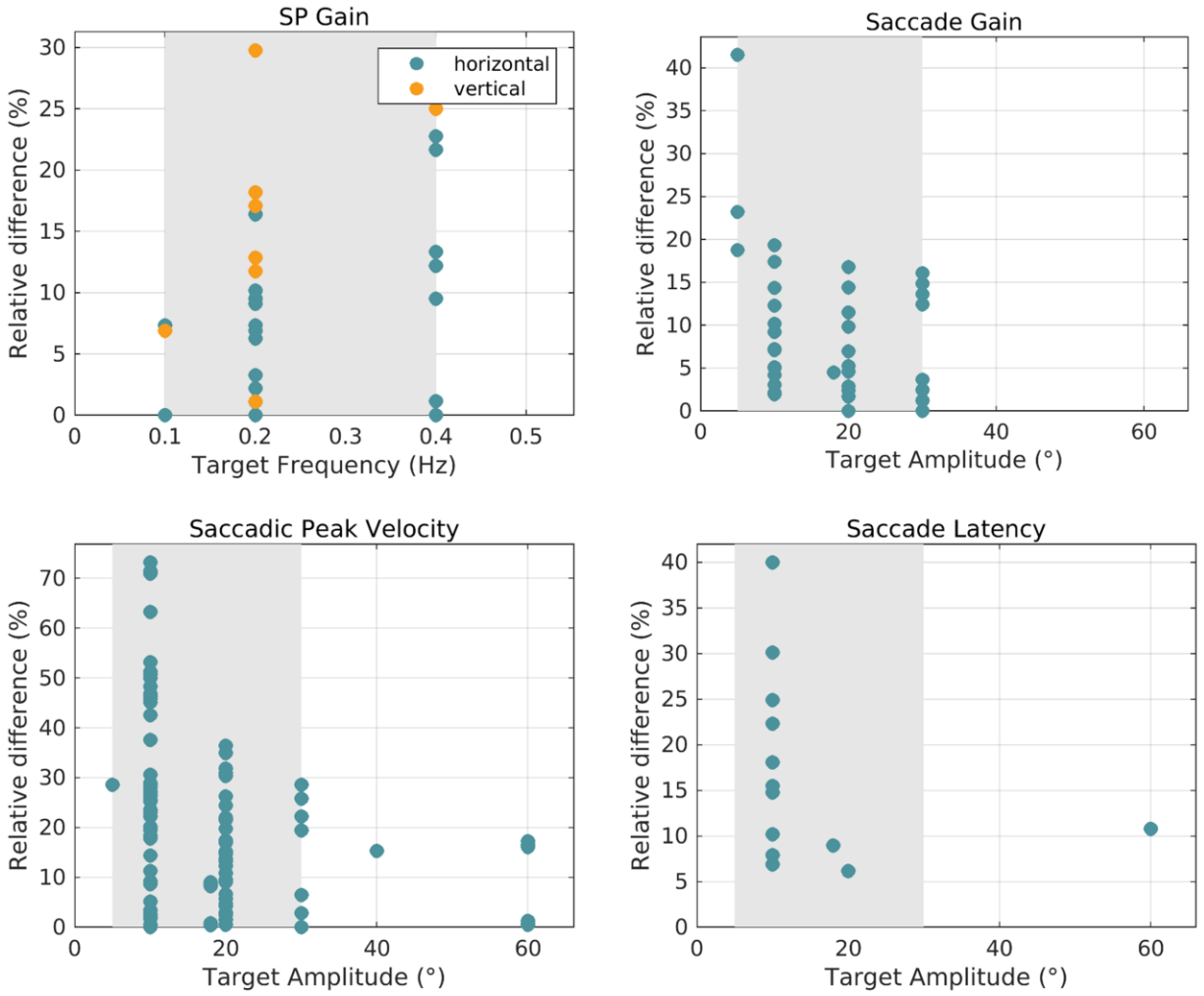


Figure S5: Variability across studies in the mean normative value. For each peak target frequency (smooth pursuit) or target amplitude (saccades), we calculated the difference between the average normative value (or average across the healthy volunteer group) for each pair of studies. This difference between studies is indicated as relative error, in percentage. The ranges of recommended target frequencies and target jump amplitudes are highlighted in gray.



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