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INVITED REVIEW

# What assessments are currently used to investigate and diagnose cerebral visual impairment (CVI) in children? A systematic review

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**Keywords:** cerebral visual impairment, childhood visual impairment, children, cortical visual impairment, diagnosis, investigation

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## Abstract

**Purpose:** Cerebral visual impairment (CVI) is the leading cause of childhood visual impairment in the developed world. Despite this, there are no agreed clinical guidelines for the investigation and diagnosis of the condition. Before development of such guidelines can commence, it is important to recognise which approaches are currently employed. This systematic review evaluated the literature to identify which methods of assessment are currently used to investigate and diagnose childhood CVI.

**Methods:** Medline, Embase, CINAHL, Scopus and the Cochrane Library databases were systematically searched in January 2020 using defined search terms. Articles were included if they: (i) were research papers, conference abstracts or research protocols published in peer-reviewed scientific journals, or relevant textbooks; (ii) included a clinical investigation of CVI in children; (iii) provided an explanation or criteria to diagnose CVI and (iv) were specifically investigating cerebral/cortical visual impairment. Methods used to a) assess and b) diagnose CVI were extracted from included articles. ‘Assessment scores’ were assigned for each method employed by researchers to investigate and diagnose CVI to quantify and compare approaches between articles. A quality grading was also applied to each article.

**Results:** Of 6454 identified articles, 45 met the inclusion criteria. From these, 10 categories of assessment utilised within included articles were identified: (1) Medical history, (2) Vision assessment/ophthalmologic examination, (3) Neuroimaging, (4) Visual behaviour and direct observation, (5) Structured history-taking, (6) Visual perception tests, (7) Ocular movement and posture assessment, (8) Intelligence/IQ assessment, (9) Clinical electrophysiology and (10) Neurodevelopmental tests. In terms of diagnostic criteria, the most commonly reported approach was one of exclusion, i.e., CVI was diagnosed when visual dysfunction could not be attributed to abnormalities detected in the anterior visual pathway.

**Conclusion:** There is a lack of common practice in the approaches used by clinicians to investigate and diagnose CVI in children. At present, a ‘diagnosis of exclusion’ remains the most common means to diagnose CVI. Development of clinical guidelines for assessment and diagnosis are necessary to ensure consistency in the diagnosis of CVI and the timely implementation of support to alleviate the impact of CVI on the child’s daily living.

## Introduction

In recent years there has been increasing interest in cerebral visual impairment (CVI) in children. CVI is a condition in which damage to the retro-chiasmatic visual pathways results in the disruption of normal visual processing.<sup>1</sup> While severe cases of CVI (previously called 'cortical blindness') were considered uncommon, CVI is now considered the most common cause of childhood visual impairment in the developed world.<sup>2–4</sup> It arises from conditions which cause abnormal development of, or damage to, the brain, affecting the visual pathways and disrupting normal visual function. Conditions leading to CVI often occur perinatally, with the most common cause reported as hypoxic-ischaemic injury.<sup>1,5–7</sup> CVI is also frequently reported among prematurely born children as their prematurity results in increased risk of insult to the developing brain.<sup>8</sup> Advances in medical care have resulted in an increased survival rate among extremely premature and low birthweight neonates and this is likely to have contributed to the increased prevalence of CVI. As such, interest and awareness of CVI among healthcare professionals is growing. Children with CVI can present with an array of visual difficulties dependent on the location and extent of damage or malformation of the brain. Damage affecting specific parts of the retro-chiasmatic visual pathways can lead to characteristic behaviours associated with CVI, for example difficulty interpreting visual information in a crowded environment, or recognising familiar objects.<sup>1</sup>

The growing recognition of the existence and relevance of CVI has led to debate about how CVI is defined,<sup>9</sup> diagnostic criteria and who can and should diagnose CVI.<sup>6</sup> A recent systematic review by Sakki *et al.*<sup>9</sup> carried out a thematic analysis of currently used definitions of childhood CVI. This review defined CVI as 'a verifiable visual dysfunction which cannot be attributed to disorders of the anterior visual pathways or any potentially co-occurring ocular impairment'. Uncertainties around diagnosis delay or prevent a child receiving the support they require at home and at school. Whilst there is no 'cure', the impact of CVI on daily living activities can be alleviated by the adoption of practical strategies and modification of the child's environment; targeting specific difficulties with which the child presents.<sup>1,10</sup> Additionally, providing a diagnosis offers parents and carers an explanation for the child's visual strengths, limitations and behaviours.<sup>8</sup> Huo *et al.*<sup>11</sup> found that children who were diagnosed with CVI before 3 years of age had an improved visual prognosis compared to children receiving a later diagnosis. Huo *et al.*'s report supports findings from other studies that show an improvement in visual function in children provided with early training and habilitation programmes.<sup>12–14</sup>

One of the challenges in assessing and diagnosing CVI is that young children, and those with the learning and/or physical disabilities that commonly coexist with CVI, are often unable to undertake the plethora of tests that aid identification of CVI. Responding to this challenge, researchers across the globe have developed a range of accessible assessments to evidence and diagnose CVI in children. These include quantitative and qualitative assessments using behavioural, clinical and visual metrics. Diagnosis of CVI generally requires a range of assessments, applied by a multi-disciplinary team of professionals and collated to create a comprehensive picture of the child's difficulties, form a diagnosis and devise a habilitation plan.<sup>15</sup> In order to determine which assessments are the most useful components of diagnostic guidelines, it is first valuable to appreciate which tools are currently utilised in the assessment and diagnosis of CVI.

The aim of this systematic review is to identify and evaluate the assessments, which are currently used to investigate and diagnose CVI in children, as determined through examination of the peer-reviewed scientific literature. A secondary aim is to determine which professionals are most often involved in assessment and diagnosis of CVI.

## Methods

### Protocol and registration

The methods used in this review were designed according to the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA).<sup>16</sup> The review protocol was registered with the International Prospective Register for Systematic Reviews online in November 2016 (registration number CRD42016051262), accessed online at <https://www.crd.york.ac.uk/prospero/>.

### Eligibility criteria

This review focused on methods and tools used to assess and diagnose CVI in children. Articles were included if they: (i) were original research papers, conference abstracts or research protocols published in peer-reviewed scientific journals, or relevant textbooks; (ii) included a clinical investigation of CVI in children (0–18 years); (iii) provided an explanation or criteria to diagnose CVI and (iv) were specifically investigating cerebral/cortical visual impairment rather than visual perception or dorsal/ventral stream function. No restrictions were placed on date of publication, sample size, gender, race or study locations. Review articles were excluded but citation lists of these articles were searched for additional papers that met the inclusion criteria.

### Search strategy

Literature searches were carried out in January 2020 by one author (ELM), following development of a search strategy by all authors and subject librarians at Ulster University, using the following databases: Medline, Embase, CINAHL, Scopus and the Cochrane Library. An example of the search terms used in Medline is included in Appendix S1. Results from database searches were stored in RefWorks where duplicates were removed. Searches were limited to English language.

### Article selection

Titles and abstracts were independently screened for suitability by two authors (ELM and JAL). Disagreements between reviewers were resolved through discussion and reference to article eligibility criteria. Full texts of articles, which met the inclusion criteria following title and abstract screening, were obtained and reviewed by ELM for eligibility. Second reviewer JAL screened 10 percent of articles included/excluded by ELM to evaluate repeatability of decisions. Where a full text was unavailable, or there was insufficient detail included in a conference abstract, attempt was made to contact the author of the publication. Manual screening of textbooks and grey literature was also carried out to identify relevant literature and determine eligibility.

### Data extraction

Data were extracted from articles that met inclusion criteria following full-text review. A data extraction tool was designed to gather publication characteristics (e.g., sample size, study aim), participant details, information on the type of tests and methods used during the CVI assessment, CVI diagnostic criteria, professionals involved in the assessment process and main findings.

### Data analysis

Initial analysis determined, for each article, which assessment tools were used to assess children with diagnosed or suspected CVI. Further analysis recorded, where available, the specific diagnostic criteria or description used by the researchers to form a CVI diagnosis. The professionals and disciplines involved in the assessment and diagnostic process were recorded.

### Quality assessment

As this review aimed to identify and evaluate the tools used to investigate and diagnose CVI in children, the quality of

articles was graded according to the detail provided on how a CVI diagnosis was achieved. Articulation of the professionals involved in the assessment was also considered, to address the secondary aim of the review. A quality assessment tool was developed for this purpose. Currently available tools were considered and deemed inappropriate for the present review as they are designed for use with randomised and non-randomised studies.<sup>17,18</sup> The assessment tool used a simple three point grading system, similar to that used by a previous review,<sup>9</sup> and graded the quality of information as 'Complete' (Grade A), 'Partial' (Grade B) and 'Incomplete' (Grade C). To achieve a grade of 'Complete' articles were required to include an explicit diagnostic criterion, for example "CVI was diagnosed...", a description of the professionals involved in the assessment and a clear description of the tests used to assess CVI. 'Partial' grades were attributed when there was a description of how the diagnosis was made, but no information on which professional(s) undertook the assessment, or if a description of how a diagnosis was made and the professionals involved were included, but the description of tests and assessments used was brief/ambiguous. An article was graded 'Incomplete' if CVI diagnosis was mentioned, but the method for reaching a diagnosis was unclear or unavailable, or little detail was provided on the assessments used to form the diagnosis. An article graded 'Incomplete' also failed to document the professionals involved in the assessment.

### Scoring of assessments

In addition to assigning a quality grade to each article as described above, an additional numerical score was attributed to each article based on which tests were used by the authors in their assessment. The purpose of this score was to quantify the scope and depth of the CVI assessment. Scores were attributed based on information available in the literature evidencing the validity of tests and assessments used. A score of 0, 1 or 2 for each assessment was possible, with a higher score equating to a more robust/well-established assessment method. The rationale for assigning scores is discussed at the end of each assessment category. For all, a score of '0' was assigned if an assessment was not undertaken. A total score of 20 was possible. A higher overall score does not necessarily equate to a better assessment, but rather allows for a more in-depth evaluation of the methods used in each article. While some categories of assessment identified in this review may be considered more appropriate when considering a diagnosis of CVI, an equal weighting of assessment score was attributed for each category in order to prevent the introduction of bias, where the authors may have attributed a higher score to a category that they considered more critical in a

CVI assessment. As is evident from Appendix S2, a wide range of professionals are involved in the assessment and diagnosis of CVI; therefore, what may be considered an important test for one group of professionals may be less important to another group.

**Results**

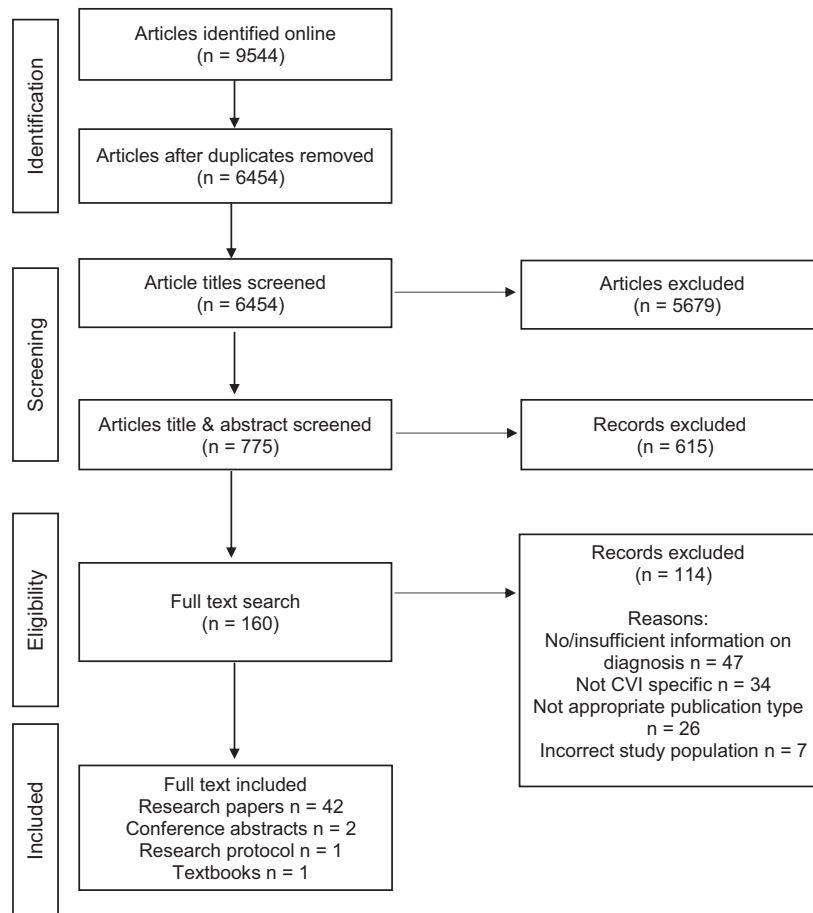
Figure 1 shows the PRISMA diagram of the article screening and review process. Cohen’s kappa was carried out to determine the level of agreement between articles double-screened for eligibility which indicated good agreement between reviewers<sup>19</sup> ( $k = 0.80, p = 0.005$ ). Forty-five articles and one textbook met the inclusion criteria outlined above. An attempt was made to contact authors of three articles for additional information. One author did not respond, contact details could not be obtained for another and data were unavailable for one article. Using the quality assessment described above, 12 articles were graded ‘Complete’, 14 ‘Partial’ and 20 were graded ‘Incomplete’.

**Tests used in assessment of participants**

All articles were reviewed and methods of assessment recorded and scrutinised. These were grouped into 10 ‘categories of assessment’: (1) Medical history, (2) Vision assessment/ophthalmologic examination, (3) Neuroimaging, (4) Visual behaviour and direct observation, (5) Structured history-taking, (6) Visual perception tests, (7) Ocular movement and posture assessment, (8) Intelligence/IQ assessment, (9) Clinical electrophysiology and (10) Neurodevelopmental test(s). Table 1 summarises and quantifies the number of categories of assessment reported in the included articles.

*Medical history*

CVI is associated with conditions or co-morbidities that may cause damage to or abnormal development of the brain. Such conditions or co-morbidities include cerebral haemorrhage, hydrocephalus, neonatal hypoglycaemia, central nervous system infections, traumatic brain injury, metabolic disorders, cerebral palsy and hypoxic-ischaemic



**Figure 1.** Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) diagram of selection process for relevant articles in review.

**Table 1.** Categories of assessment reported in included articles, where X = use reported by article and - = use not reported by article for each assessment category

Articles	Quality assessment	Participant details			Assessment categories									
		Mean age (years)	Age range (years)	Sample size	Medical history	Vision assessment/ Ophthalmologic examination	Visual behaviour and direction observation	Structured history -taking	Visual perceptual and posture tests	Ocular movements	Intelligence test	Clinical electrophysiology	Neuro-developmental assessment	Number of assessment methods used
Philip (2017)	A			1478	X	X	X	X	-	X	-	-	-	7
Duke et al. (2019)	A		4–16	370	X	X	X	X	X	-	-	-	-	7
Bosch et al. (2014)	A	(Mdn 3)		309	X	X	X	-	-	X	X	-	-	6
Andersson et al. (2006)	A	(Mdn 9.3)		140	X	X	X	X	-	X	-	X	-	7
Handa et al. (2018)	A	1.53	0.24–6.37	53	X	X	X	-	-	X	X	-	-	4
Whiting et al. (1985)	A		0.5–19	50	X	X	X	-	-	X	X	-	-	6
Oribus et al. (2011)	A	6.83	3.42–17	45	X	X	X	X	X	X	-	-	-	8
van Genderen et al. (2012)	A	8	5–16	30	X	X	X	X	X	X	-	-	-	6
Oribus et al. (2009)	A	0.5	4–20	70	X	X	X	X	X	X	-	-	-	5
Ferziger et al. (2011)	A	8.25	3–20	51	X	X	X	X	-	X	-	-	-	6
Geldof et al. (2015)	A	5.5		25	X	X	X	X	X	X	-	-	-	7
Roman-Lantzy (2007)	A				X	X	X	X	-	-	-	-	-	3
Matsuba & Jan (2006)	B			423	X	X	X	X	-	X	-	-	-	3
Ben Itzhak et al. (2019)	B	(Mdn 6.42)	3–20	179	X	X	X	X	X	X	-	-	-	6
Jasper & Philip (2018)	B	4 definite CVI group, 2 probable CVI group	0.25–17	167	X	X	X	X	-	-	X	-	-	7
Fazzi et al. (2007)	B	4.5	0.25–15	121	X	X	X	X	X	X	X	X	X	9
Brodsky et al. (2002)	B			100	X	X	X	X	-	X	-	-	-	4
Khetpal et al. (2007)	B	3.1	0.2–19	98	X	X	X	X	X	X	-	-	-	5
Mitry et al. (2016)	B		4–15	90	X	X	X	X	X	X	-	-	-	4

(continued)

Table 1. (continued)

Articles	Participant details			Quality assessment (years)	Age range (years)	No. of CVI controls	No. of controls	Assessment methods used									
	Mean age (years)	Sample size						Vision assessment/ Ophthalmologic examination	Neuro-imaging	Visual behaviour and direction observation	Structured history-taking	Visual perceptual tests	Ocular movements and posture	Intelligence test	Clinical electrophysiology	Neuro-developmental assessment	Number of assessment methods used
Salati et al. (2003)	7.1	56	56	X	2-16		X	X	X	X	X	X	X	X	X	8	
Kooiker et al. (2015)	8.5	56	48	X	1.08-12.9		X	X	X	X	X	X	X	X	X	5	
Macintyre-Beon et al. (2013)		130	46	X	5.5-12.3		X	X	X	X	X	X	X	X	X	7	
Skoczinski & Good (2004)	3.5	35	35	X	0.33-16		X	X	X	X	X	X	X	X	X	4	
Weinstein et al. (2012)	6.75	81	19	X	5-16		X	X	X	X	X	X	X	X	X	5	
Vandeleef et al. (2020a)	6.83	25	12	X	4-9.08		X	X	X	X	X	X	X	X	X	5	
Salati et al. (2001)	5	11	11	X	1-9		X	X	X	X	X	X	X	X	X	6	
Philip et al. (2016)	3.8	342	342	X	0-17		X	X	X	X	X	X	X	X	X	5	
Peherer et al. (2018)	5.24	124	124	X			X	X	X	X	X	X	X	X	X	4	
Huo et al. (1999)	3	170	170	X			X	X	X	X	X	X	X	X	X	4	
Hård et al. (2004)	(Mdn 6.7)	91	91	X			X	X	X	X	X	X	X	X	X	6	
Dutton et al. (1996)		90	90	X			X	X	X	X	X	X	X	X	X	3	
Salavati et al. (2015)	9.5	77	77	X	4.17-12		X	X	X	X	X	X	X	X	X	4	
Houliston et al. (1999)		200	52	X			X	X	X	X	X	X	X	X	X	4	
Cioni et al. (1997)	11.8	48	48	X			X	X	X	X	X	X	X	X	X	5	
Cioni et al. (1996)	(Mdn 2.21)	48	48	X			X	X	X	X	X	X	X	X	X	6	
Kooiker et al. (2014)	7.3	127	42	X	1.1-12.9		X	X	X	X	X	X	X	X	X	5	
Good (2001)		41	41	X	0.5-16		X	X	X	X	X	X	X	X	X	4	
Salavati et al. (2017)	9.4	37	37	X	4.5-12		X	X	X	X	X	X	X	X	X	3	
Macintyre-Beon et al. (2012)	10.8	36	36	X	5-16.5		X	X	X	X	X	X	X	X	X	1	

(continued)

Table 1. (continued)

Articles	Quality assessment	Participant details			Sample size										
		Mean age (years)	Age range (years)	No. of CVI	Medical history	Vision assessment/ Ophthalmologic examination	Neuro-imaging	Visual behaviour and direction observation	Structured history-taking	Visual perceptual tests	Ocular movements and posture	Intelligence test	Clinical electrophysiology	Neuro-developmental assessment	Number of assessment methods used
Good <i>et al.</i> (2012)	C	1.94	0.42–5	34	16	X	X	-	-	-	-	X	-	-	4
Chen <i>et al.</i> (1992)	C			30		X	X	-	-	X	-	-	X	-	5
Lanzi <i>et al.</i> (1998)	C	3.5	1.67–5.5	23	12	X	X	-	-	X	-	X	X	-	6
Franki <i>et al.</i> (2017)	C	4.25		23	51	-	-	-	-	X	-	-	-	-	2
Weiss <i>et al.</i> (2001)	C	0.51	0.08–1.08	17	31	X	X	-	-	-	X	X	X	X	6
Eken <i>et al.</i> (1996)	C		0.77–1.5	9		X	X	X	-	-	X	X	X	X	7
Suner <i>et al.</i> (2016)	C					-	-	-	-	-	-	-	-	-	1
Total number of articles						43 (93.5%)	43 (93.5%)	29 (63%)	19 (41%)	17 (37%)	12 (26%)	33 (72%)	16 (35%)	15 (33%)	8 (17%)

Quality assessment: A = Complete, B = Partial, C = Incomplete used to order articles, followed by number of participants with CVI included in article. Mdn = median.



encephalopathy. The latter is the most common cause of CVI.<sup>1,20–23</sup> Consideration of the child's medical history in the assessment and diagnostic process adds value in helping to identify children who are most 'at-risk' of CVI.<sup>24</sup>

In the present review, details of the child's medical history or diagnoses were documented in 43 articles (93.5%). Articles that clearly documented the children's medical history and diagnoses with sufficient detail were assigned a score of 2. Articles which reported more general information on the study sample's medical history (for example, the study was carried out in a population of children with cerebral palsy, but no further information on the study sample was provided), or reported that medical history was accessed through medical notes, but did not provide further detail on this, scored 1. Articles that did report on the children's medical history scored 0.

#### *Vision assessment/ophthalmologic examination*

Assessment of ocular health and visual function is vital when examining children with suspected CVI. Visual deficits must be identified and managed to rule out ocular causes of visual impairment that may account for the child's visual difficulties. Vision assessment, including visual acuity measurement as a minimum, was reported in 43 articles (93.5%). Of three articles which did not include information on vision assessments, one was a conference abstract, and although it is likely that vision tests were carried out as part of the study, this information was not reported in the published abstract.<sup>25</sup> Another was a textbook which detailed an approach to CVI assessment which included having a 'normal or near normal eye examination that cannot explain the child's impaired vision'; however, a description of the tests used to determine the normality (or otherwise) identified by the eye examination were not documented.<sup>26</sup> The remaining article was a short report discussing validation of the visual skills inventory questionnaire.<sup>27</sup> All other articles reported a visual acuity measurement, 20 (43.5%) reported measuring the participants' refractive status,<sup>11,20,21,28–44</sup> 26 (56.5%) documented that a visual field assessment was conducted<sup>7,11,21,28,31,32,35–38,40–42,44–56</sup> and 26 (56.5%) reported assessment of ocular health.<sup>7,11,20–22,28–36,40–44,46,48,51,57–60</sup> Five (11.1%) articles documented assessment of contrast sensitivity,<sup>21,37,38,42,61</sup> seven (15.6%) reported measuring stereopsis<sup>21,35,37,38,42,48,49</sup> and four (8.9%) articles measured accommodative accuracy.<sup>32,40,42,49</sup>

For vision assessment/ophthalmological examination, a score of 2 was assigned if an article reported assessment of refractive error, ocular health and visual acuity using a validated and/or well-established test. A score of 1 was assigned if an article reported assessment of at least one aspect of visual assessment, i.e., refractive error, ocular health or visual acuity (using any method to assess visual acuity).

Remaining articles who did not report any form of vision assessment were scored 0.

#### *Neuroimaging*

Neuroimaging is a central tool used in the detection and diagnosis of pathology in the brain.<sup>62</sup> Three techniques are commonly used to image the infant brain: ultrasound, computed tomography (CT) and magnetic resonance imaging (MRI). Thukral<sup>62</sup> reviewed the problems and preferences in paediatric brain imaging and reported that ultrasound is the preferred technique for screening, MRI is best used for investigating brain tissue and anatomy, and that CT is reserved for trauma evaluation. Blankenberg *et al.*<sup>63</sup> further support the use of MRI or CT rather than ultrasound to aid the diagnosis of paediatric neurological problems.

Neuroimaging was reported in 29 (63.0%) articles. The most commonly reported imaging technique was MRI ( $n = 22$ ).<sup>20,21,28–34,41,44,45,47,48,50,51,56–58,64–66</sup> Ten articles reported use of CT scans<sup>20,21,29–31,35,46,51,58,65</sup> and five reported use of ultrasound.<sup>29,31,45,48,51</sup> Ten articles reported use of more than one neuroimaging modality.<sup>20,21,29–31,45,48,48,51,65</sup> Of the publications that included neuroimaging results, the majority ( $n = 23$ , 82.1%) were obtained retrospectively from the child's medical records. Four of the remaining six articles reported use of MRI at the time of assessment;<sup>21,34,47,57</sup> one reported a mixture of all three neuroimaging modalities<sup>29</sup> and one did not specify the type of neuroimaging technique used.<sup>42</sup> This resulted in a considerable amount of missing neuroimaging data and a lack of consistency in the type of neuroimaging assessment undertaken i.e., some participants may have had a CT scan, while others had a MRI scan. Five articles did not report which neuroimaging technique had been used, but simply stated that neurological assessments were performed or results were available from medical notes.<sup>11,42,49,67,68</sup>

For neuroimaging, a score of 2 was assigned if neuroimaging was carried out contemporaneously at the time of assessment and a score of 1 was assigned if results were obtained retrospectively from medical records. Articles that did not report use of neuroimaging were scored 0.

#### *Visual behaviour and direct observations*

An assessment of visual behaviour through direct observation can provide valuable information regarding a child's visual strengths and weaknesses. Such observations can help identify a child's key challenges and allow targeted interventions to be introduced in order to assist the child's daily living.<sup>69</sup> Nineteen articles (41.3%) reported observation of the child's visual behaviour as part of the CVI assessment. Included in the evaluation of visual behaviour was observation of blink reflex ( $n = 1$ ),<sup>29</sup> visual threat response ( $n = 2$ ),<sup>29,42</sup> interaction with objects ( $n = 1$ ),<sup>57</sup> visual

environmental exploration ( $n = 3$ ),<sup>34,42,57</sup> photophobia ( $n = 1$ ),<sup>20</sup> visual fixation ( $n = 2$ ),<sup>42,44</sup> visual attention ( $n = 4$ )<sup>37,38,42,50</sup> and light perception ( $n = 2$ ).<sup>32,57</sup> Three articles reported observation of the child's spontaneous visual behaviour,<sup>21,49,51</sup> while another reported ophthalmological observation of the child.<sup>60</sup> One article reported clinical observation in accordance with Huo's criteria,<sup>11,22</sup> and two articles reported children underwent 'clinical observation' but did not detail what this entailed.<sup>41,54</sup> Roman-Lantzy advocates for observing a child's visual behaviour in a range of settings, in addition to presenting the child with different visual stimuli during the clinical assessment in order to assess how the child responds or interacts with these stimuli.<sup>70</sup>

If an article reported specific detail on which behaviours were observed, and noted a minimum of two behaviours, a score of 2 was assigned. Where an article stated observations of visual behaviour were carried out, but provided no information on which behaviours were seen, or reported on one behaviour only, a score of 1 was assigned. Articles that did not report any assessment of visual behaviour scored 0.

#### *Structured history-taking*

Opinions of parents and carers involved in a child's care on a daily basis allows unique insight into the child's habitual visual and behavioural strengths and limitations.<sup>71</sup> By contrast, clinicians are only able to observe the child for a short time in an unfamiliar environment. While the in-clinic assessment provides valuable information, it is unlikely to reveal the true extent of the child's difficulties in all situations. As such, parental interview and questioning affords valuable, additional insight into the child's visual function.

Seventeen articles (37.0%) reported the use of structured history-taking to explore the child's visual behaviours as a way of assessing functional vision. History-taking was primarily conducted using a clinician-administered questionnaire directed at the parent or carer at the time of assessment. This method allows clarification and further exploration of any reported vision difficulties. Thirteen articles reported use of a version of the Visual Skills Inventory (VSI) which was developed based on difficulties observed and reported by Dutton and colleagues in 1996.<sup>27,28,31,35-39,41,42,49,53,60</sup> The first iteration of this questionnaire contained 22 questions.<sup>36</sup> Various adaptations have been made to the VSI following its initial development. Macintyre-Beon *et al.*<sup>37</sup> used the questionnaire to explore CVI behaviours in a population of prematurely born children, using an extended 48-question version of the original questionnaire. A further two articles included in the present review reported use of a 52-item version, referred to as the INSIGHT questionnaire.<sup>39,42</sup>

Roman-Lantzy advocates for parent/carer input into the assessment and diagnosis of CVI, and suggests using parent, carer or educator interview to elicit evidence of characteristic behaviours associated with CVI. In her book, she uses a 25-item questionnaire which parents complete during a face-to-face interview with the clinician.<sup>72</sup> Two articles included in the present review reported use of the Flemish CVI questionnaire developed by Ortibus and colleagues.<sup>50,66</sup> This questionnaire was designed as a screening tool to seek evidence of behaviours associated with CVI and contains 46 items.<sup>50</sup> Furthermore, Ferziger *et al.*<sup>32</sup> developed a 26 item functional vision questionnaire for completion by the child's primary educator designed to assess children's daily visual performance.

Other questionnaires used in the present review to explore the child's behaviour include the Strengths and Difficulties Questionnaire (SDQ),<sup>38,42</sup> Children's Social Behaviour Questionnaire (CSBQ)<sup>38</sup> and the Pediatric Quality of Life Inventory (PedsQL).<sup>39,42</sup>

Articles which reported use of previously published questionnaires that explored CVI specific behaviours were assigned a score of 2 and questionnaires which were not CVI specific scored 1. Articles that did not report any structured history-taking were scored 0.

#### *Visual perception tests*

Children with CVI often present with visual perceptual difficulties. A wide range of tests are available to examine aspects of visual perception, e.g., tests of visual memory and attention.<sup>39,73-76</sup> Use of tests to measure various aspects of visual perception were reported in 12 articles (26.1%) in the present review. The L94 visual perception battery<sup>47</sup> was the most frequently reported ( $n = 4$ ).<sup>47,50,56,63</sup> Use of the Test of Visual Perceptual Skills-Revised (TVPS-R) was reported in two articles,<sup>31,50</sup> as was the Developmental Test of Visual Perception (DTVP).<sup>21,37</sup> The Stirling Face Processing test,<sup>37</sup> LEA 3D puzzle<sup>42</sup> and Heidi expression facial recognition test<sup>42</sup> were each reported in one article. Use of the Beery Visual-Motor Integration test (VMI),<sup>50,56</sup> LEA mailbox<sup>39,42</sup> and LEA rectangles<sup>39,42</sup> were each reported in two articles. A child was required to pick objects up from a patterned and plain background in two articles.<sup>39,42</sup> Global form and motion visual coherence were each assessed by a single article.<sup>37,38</sup> The Child Visual Impairment Test for 3- to 6-year-olds (CVIT 3-6) was employed in one article.<sup>56</sup> Two articles did not report which tests were used; one of these reported that a neuropsychological test battery was used to assess visual perception but a description of the test was not provided.<sup>28</sup> One article reported that information regarding visual perceptual ability was extracted from the child's medical records; however no information on how the ability had been measured was reported.<sup>55</sup>

The L94 battery comprises eight tasks which collectively assess ability to identify everyday objects, visuo-constructive ability and form discrimination.<sup>77</sup> The TVPS-R consists of seven subscales which measure visual discrimination, visual memory, visual spatial relationships, visual form constancy, visual sequential memory, visual figure ground and visual closure.<sup>78</sup> The DTVP has five subscales which are designed to measure motor-enhanced visual perception and motor reduced visual perception, in addition to an overall general visual perception index.<sup>79</sup> The VMI consists of three tasks; one core, and two supplementary. The core task assesses the integration of visual perception and motor skills, while the supplementary tasks assess visual ability without the integration of fine motor skills and fine motor skills when not integrated with visual perceptual ability.<sup>80</sup> The Stirling Face Processing Test measures a child's ability to identify and match faces.<sup>81</sup> The LEA Mailbox and LEA rectangles tests assess the perception of line direction and the length of lines respectively.<sup>82</sup> The LEA 3D puzzle task assesses visual guidance of movement and visual memory, and the HEIDI Expressions Test assesses a child's ability to interpret facial expressions.<sup>42</sup> The CVIT-3-6 is a recently developed tool designed for assessing the broad range of visual perception impairments which are commonly reported in children with CVI.<sup>83</sup>

Each of these tests assess different aspects of visual perception. Whilst acknowledging that articles included in the present review did not consistently assess the same facets of visual perception, a score of 2 was assigned if the article described the visual perceptual measure employed. Where authors did not provide this information a score of 1 was assigned. A score of 0 was assigned if an article did not report carrying out an assessment of visual perception.

#### *Ocular movements and posture*

Children with CVI often present with oculomotor deficits<sup>1,20</sup> including nystagmus,<sup>20,21</sup> strabismus<sup>20,84</sup> and abnormal saccadic and smooth pursuit eye movements.<sup>1,21</sup> Thirty-two articles (69.6%) reported carrying out an ocular movement assessment<sup>7,11,20,21,29–38,40–46,55,57–60,65,68</sup> and 26 (56.5%) an ocular posture/alignment assessment.<sup>11,20,21,29–38,40–45,48–50,55,58,59,66</sup> Of those who specified details of the ocular movement assessment, six included saccadic eye movement assessment,<sup>21,34,36,42,49,66</sup> four smooth pursuits,<sup>21,42,57,65</sup> nine assessed the optokinetic nystagmus response (OKN),<sup>21,32,41,45,46,49,51,57,65</sup> eight assessed the child's ability to fix and follow,<sup>21,29,32,34,44,49,50,57</sup> and two assessed convergence.<sup>38,42</sup> Twenty-three articles commented on whether the child presented with nystagmus eye movements.<sup>7,11,20,21,29–36,38,40,42,44,50,55,58,59,66,68</sup>

If an article detailed the method of assessing and/or results of an ocular movement and posture assessment a

score of 2 was assigned. Articles that detailed ocular movement or posture assessment, or OKN only, scored 1. Articles that did not report assessment of ocular movements, or reported only whether the child had nystagmus or not, scored 0.

#### *Intelligence/cognitive (IQ) assessment*

Intelligence tests are often used to provide an overall assessment of general cognitive functioning.<sup>85</sup> Two commonly used intelligence tests are the Wechsler Intelligence Scale for Children (WISC), designed for use in children aged 6 years and over, and the Wechsler Preschool and Primary Scale of Intelligence (WPPSI), used for children aged 2.6–7.7 years. The WISC tests a child's verbal comprehension, perceptual reasoning, working memory and processing speed.<sup>86</sup> The WPPSI measures full scale IQ, verbal comprehension, working memory, visual spatial index, fluid reasoning and processing speed.<sup>87</sup>

An alternative intelligence test is the Snijders-Oomen Non-verbal Intelligence test which can be used with very young children and those with poor communication and language development.<sup>88</sup> The Snijders-Oomen tests a child's abstract reasoning, concrete reasoning, spatial and perceptual tests.<sup>89</sup> The Kaufman Brief Intelligence test is another readily available IQ test that includes both verbal and non-verbal scales which collectively assess expressive vocabulary, verbal knowledge and matrices. The non-verbal matrices subtest aims to measure fluid reasoning and visual processing.<sup>90,91</sup> The developers of the test suggest it should be used for screening rather than diagnostic purposes.

In this review, 16 articles (34.8%) reported results from IQ assessment. Seven recorded that this information was extracted from medical notes,<sup>49,52,54–56,66,68</sup> or if not available, testing was conducted during the assessment.<sup>56</sup> Three articles which extracted results of IQ assessment from medical notes did not list which tests were used to ascertain intelligence. The most common intelligence test employed by included articles was the Wechsler Intelligence Scale ( $n = 8$ ).<sup>21,28,34,35,38,47,50,66</sup> Two articles classified intelligence according to the Committee of Test Affairs in Netherlands (COTAN) criteria for IQ scores; however the authors did not state which tests were used to obtain the scores.<sup>52,55</sup> Four articles reported use of the Snijders-Oomen non-verbal intelligence test,<sup>47,50,56,66</sup> and one article used the Kaufman Brief Intelligence Test (KBIT).<sup>37</sup> The remaining three articles did not state which tests were carried out.<sup>46,49,54</sup>

Considering this information, articles that documented use of a well-established, recognised test scored 2, and those who did not list which tests were used in their assessment scored 1. If IQ assessment was not carried out, an article scored 0 in this section.

### *Clinical electrophysiology: electroretinography, visual evoked potentials and electroencephalography*

Electrophysiology is used to measure the function of living tissue using electrical and chemical signals.<sup>92</sup> Fifteen articles (32.6%) reported use of at least one clinical electrophysiology method to measure brain or visual function in response to visual stimuli. Eleven of these reported use of visual evoked potentials (VEPs),<sup>21,22,29,33,46,48,51,57,59,65,67</sup> five articles used electroretinography (ERG)<sup>33,43,44,46,57</sup> and seven used electroencephalography (EEG).<sup>21,22,33,34,41,46,51</sup> Each of these electrophysiology techniques measure different functions; ERGs assess the functional integrity of the retina,<sup>93</sup> VEPs are used to determine the integrity of the visual pathways from the macula to the visual cortex<sup>94</sup> and EEG is used in the diagnosis of neurological disease and to monitor brain activity.<sup>95</sup>

A score of 2 was assigned to articles that reported use of VEP, as this provides relevant information on the function of the primary visual pathway. EEG and ERG were both assigned a score of 1 as these measures provide less specific information in terms of a child's visual function. Articles that did not report use of clinical electrophysiology testing were scored 0 for this section.

### *Neurodevelopmental assessment*

It is recommended that children who are at risk of developmental delay, for example, children who are born preterm or have suffered hypoxic insult at birth, undergo developmental testing to assess mental and psychomotor development.<sup>96</sup> Many of these children at risk of developmental delay are also at risk for CVI due to the associated aetiologies. In this review, eight articles (17.4%) reported use of a neurodevelopment test as part of their assessment. Of these, four reported use of the Griffiths Mental Development Scales only,<sup>21,33–35,97</sup> one article employed both the Dubowitz protocol<sup>98</sup> and the Griffiths developmental scales<sup>29</sup> and one used Bayley Scales of Infant and Toddler Development.<sup>65,99</sup> Two articles did not state which neurodevelopmental test was used.<sup>30,40</sup>

The Griffiths<sup>97</sup> and Bayley scales<sup>99</sup> are two commonly used instruments to measure development in infants. Several studies have compared the two scales. Cirelli et al.<sup>96</sup> conclude that while the scores between the two instruments are not interchangeable, the meaning of the results from each test are the same indicating the validity of both tests for the use in neurodevelopmental assessment. In addition, Ramsay and Fitzhardinge<sup>100</sup> contend that the Griffiths test lacks scoring precision compared with the Bayley scales. Cirelli et al.<sup>96</sup> suggest that the Bayley test is used more often in research, whereas the Griffiths may be more suited to clinical use.

Taking this information into consideration, tests which were well-established or validated were scored 2 and articles

which did not state which test was used, or employed tests which were less well-established scored 1. Articles that did not administer a neuro-developmental test scored 0.

In summary, it is apparent that the methods used to assess children with suspected CVI are manifold. Even within each category of assessment, there is little consistency in the tests applied across articles.

### **Assigned assessment utility scores**

Using the scoring system outlined above, the highest possible total score assigned based on the scope and depth of assessment methods used by the article was 20. None of the included articles applied tests covering all assessment categories, and thus, none scored 20. Total utility scores assigned to each article are shown in Appendix S2. Fazzi et al.<sup>21</sup> were awarded the highest score, having covered nine categories of assessment and achieving the highest possible score within each assessment category, with the exception of the 'visual behaviours and direct observation' category. This was closely followed by Salati et al.<sup>34</sup> with a score of 15. Salati et al.<sup>34</sup> covered eight categories of assessment, achieving the highest possible score within each category with the exception of the 'intelligence/cognitive assessment' category.

### **Diagnostic methods for CVI**

The first section of this review discussed the assessments used to investigate CVI. The following section reviews more specifically how each article reports the means by which a CVI diagnosis was formed.

Table 2 shows how each article assigned a CVI diagnosis. Articles often provided a written description to inform the reader how a diagnosis was made, rather than detailing results of specific assessment procedures. The most commonly reported diagnostic description utilised results from the vision assessment, articulating the presence of CVI was determined where 'visual dysfunction which could not be accounted for based on ocular examination findings/anterior pathway abnormalities' ( $n = 22$ , 47.8%).<sup>7,11,20–22,29,30,32–34,40,41,43,44,48,51,52,54,58,59,65,67</sup> The presence of normal pupillary reactions was included by five articles as part of this diagnostic description.<sup>22,30,43,59,67</sup> An additional six articles also reported using results from the vision assessment/ophthalmologic examination to form a diagnosis ( $n = 28$  in total, 60.9%).<sup>28,38,56,59–61</sup> Of these, a crowding ratio derived from visual acuity measures was considered by one article<sup>28</sup> and a novel method for assessing visual function and diagnosing CVI using a computer-based system relying on the child's eye movements was reported as the only method to detect and quantify CVI by one article.<sup>61</sup>

**Table 2.** Diagnostic tests and descriptions described by included articles, where X = use reported by article, - = use not reported by article for each diagnostic category

Article	Vision measures/ ophthalmologic exam		Neurological findings		Medical history		Structured history-taking		Visual perception tests		Observation of characteristic behaviours associated with CVI		Intelligence assessment with (IQ) assessment results		Electro-physiological results		Not clear diagnosis		Number of assessment methods used (taken from Appendix S2)	Assessment utility score	Quality assessment	
	measures/ ophthalmologic exam	Neurological findings	Medical history	Structured history-taking	Visual perception tests	Observation of characteristic behaviours associated with CVI	Intelligence assessment with (IQ) assessment results	Electro-physiological results	Not clear diagnosis	Number of assessment methods used (taken from Appendix S2)	Assessment utility score	Quality assessment										
Duke et al. (2019)	-	-	-	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	7	13	A	
Geldof et al. (2015)	X	-	-	-	X	-	-	-	X	-	-	-	-	-	-	-	-	-	2	7	13	A
Ortibus et al. (2011)	X	X	X	-	X	-	-	-	X	-	-	-	-	-	-	-	-	-	4	8	13	A
Andersson et al. (2006)	-	-	-	-	-	-	-	X	-	-	-	-	-	-	-	-	-	-	1	7	11	A
Philip (2017)	X	X	X	X	-	-	-	X	-	-	X	-	-	-	-	-	-	-	6	7	10	A
van Genderen et al. (2012)	X	-	-	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	2	6	10	A
Bosch et al. (2014)	X	-	-	-	-	-	-	-	X	-	-	-	-	-	-	-	-	-	2	6	10	A
Ortibus et al. (2009)	-	-	-	-	X	-	-	-	-	-	-	-	-	-	-	-	-	-	1	5	9	A
Ferziger et al. (2011)	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	1	6	9	A
Whiting et al. (1985)	-	-	-	-	-	-	-	-	-	-	-	-	X	-	-	-	-	-	1	6	8	A
Handa et al. (2018)	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	1	4	7	A
Roman-Lantzy (2007)	-	-	-	-	-	-	-	X	-	-	-	-	-	-	-	-	-	-	2	3	5	A
Fazzi et al. (2007)	X	X	X	-	-	-	-	-	-	-	-	-	-	X	-	-	-	-	3	9	17	B
Salati et al. (2003)	X	X	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	2	8	15	B
Macintyre-Beon et al. (2013)	-	-	-	X	-	-	-	X	-	-	-	-	-	-	-	-	-	-	1	7	11	B
Jasper & Philip (2018)	X	-	X	X	-	-	-	X	-	-	-	-	-	-	-	-	-	-	3	7	11	B
Salati et al. (2001)	-	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	1	6	10	B
Ben Itzhak et al. (2019)	-	X	X	-	X	-	-	-	X	-	-	-	-	-	-	-	-	-	4	6	9	B
Weinstein et al. (2012)	X	X	X	-	-	-	-	-	X	-	-	-	-	-	-	-	-	-	2	5	8	B
Vancleef et al. (2020)	X	X	X	-	-	-	-	-	X	-	-	-	-	-	-	-	-	-	4	5	7	B
Kooiker et al. (2015)	-	-	-	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	2	5	7	B
Khetpal et al. (2007)	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	1	5	6	B
Skoczinski & Good (2004)	X	-	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	2	4	6	B
Mitry et al. (2016)	-	-	-	X	-	-	-	X	-	-	-	-	-	-	-	-	-	-	1	4	6	B
Brodsky et al. (2002)	X	X	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	2	4	6	B
Matsuba & Jan (2006)	X	X	X	X	-	-	-	-	-	-	X	-	-	-	-	-	-	-	5	3	5	B
Eken et al. (1996)	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	1	7	14	C
Hård et al. (2004)	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	6	6	10	C
Lanzi et al. (1998)	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	1	5	11	C
Weiss et al. (2001)	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	1	6	8	C
Philip et al. (2016)	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	5	5	7	C
Peherre et al. (2018)	X	X	X	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	2	4	7	C

(continued)

Table 2. (continued)

Article	Vision ophthalmologic exam		Neurological findings		Medical history		Structured history-taking		Visual perception tests		Observation of characteristic behaviours associated with CVI		Intelligence assessment		Electro-physiological results		Not clear		Quality assessment				
	measures/	exam	findings	history	history taking	tests	CVI	(IQ)	assessment	results	clear	assessment	assessment	from	Appendix	Quality	assessment	assessment	assessment	assessment			
Good (2001)	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	7	C	
Houlston et al. (1999)	-	-	-	-	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	4	C	
Cioni et al. (1996)	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	6	C	
Good et al. (2012)	X	-	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	6	C	
Chen et al. (1992)	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	6	C	
Cioni et al. (1997)	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	6	C	
Huo et al. (1999)	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	6	C	
Koölker et al. (2014)	-	-	-	X	-	-	-	-	-	-	-	X	-	-	-	-	-	-	-	-	6	C	
Dutton et al. (1996)	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	5	C	
Salavati et al. (2015)	X	-	-	X	-	-	-	-	-	-	X	-	-	-	-	-	-	-	-	-	5	C	
Salavati et al. (2017)	X	-	-	X	-	-	-	-	-	-	X	-	-	-	-	-	-	-	-	-	4	C	
Franki et al. (2017)	-	-	-	-	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	3	C	
Macintyre-Beon et al. (2012)	-	-	-	-	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	2	C	
Suner et al. (2016)	X	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	-	1	C	
Total number of articles using diagnostic method	28	13	12	9	6	6	6	5	3	4	4	1	1	1	1	1	1	1	1	1	1	1	C

Quality assessment: A = Complete, B = Partial, C = Incomplete.

The next most common method for forming a diagnosis was based on findings from neuroimaging ( $n = 13$ )<sup>7,21,34,40,48–50,52,56–58,66,67</sup> Eleven of these articles reported the use of additional methods of assessment alongside neuroimaging findings to reach a diagnosis. Consideration of the child's medical history to determine whether conditions known to be associated with CVI were present was reported as contributing to a diagnosis in 12 articles; all used medical history in conjunction with other metrics to form a diagnosis.<sup>7,28,41,49,50,52,54,55,59,66–68</sup> Structured history-taking was reported as contributing to the diagnostic process by eight articles,<sup>27,35–37,39,41,42,49</sup> with six reporting this as the only assessment used to form a diagnosis.

IQ assessment contributed to the diagnosis in five articles<sup>49,55,56,66,68</sup> and direct observation of visual behaviours was also reported as contributing to a diagnosis in five articles<sup>7,44,49,52,54</sup>; results of IQ assessment and direct observation of visual behaviours were used in conjunction with other methods of assessment.

Results from clinical electrophysiology testing were reported as contributing to a diagnosis in three articles<sup>7,21,46</sup> and one article reported that results of a 'visuo-motor assessment' were used when forming a diagnosis, however no information on what this entailed was provided.<sup>66</sup> These assessments were used in conjunction with other methods to form a diagnosis. The L94 visual perception battery was reported as the only diagnostic method used in two articles.<sup>47,64</sup> Other tests of visual perception were used by three articles,<sup>38,50,56</sup> all in conjunction with other metrics to form a diagnosis. Diagnostic criteria were unclear or not well documented in four articles.<sup>31,45,53,60</sup>

Twenty-one articles (46.7%) reported using a single method to diagnose CVI (*Table 2*). The most common of these was on the basis that visual deficits could not be accounted for based on vision assessment/ophthalmological examination findings ( $n = 11$ ) and the second most frequent single method reported was structured history-taking ( $n = 6$ ). Eleven articles reported using two measures to form a diagnosis, five used results from three assessments, three used four assessments and one article each used combinations of five or six methods to diagnose CVI (*Table 2*).

### Professionals involved in assessment

If an article specifically mentioned which professionals were involved in the assessment of CVI, this was recorded. Of the 46 articles which were included in the present analysis, 18 (39.1%) did not state who was involved in the CVI assessment and diagnostic process. Use of multidisciplinary input was documented in 18 articles (39.1%). Multidisciplinary input was recorded where input from two or more of the following disciplines was reported: medicine

(non-vision) (which includes (neuro-) paediatrician, neuro-radiologist, neurologist); vision (ophthalmologist, optometrist, orthoptist); therapy (physiotherapist, speech therapist, occupational therapist, developmental coach, therapists); psychology (neuro-psychiatrist, neuro-psychologist, psychiatrist, psychologist); and trained researchers. The authors acknowledge that the professions listed could be included in more than one category, but for the purposes of this review, professionals have been grouped into one category only as described. Multidisciplinary input was also indicated where the article stated the involvement of a multidisciplinary team, even if specific disciplines were not explicitly described ( $n = 3$ ).

The list of professionals involved in the CVI assessment, along with the number of articles which report involvement of these professionals, are listed in Appendix S3. Vision professionals were most frequently involved in the assessment of CVI ( $n = 21$ ), with ophthalmologists the most common ( $n = 19$ ). Neuro-specialists were also frequently involved ( $n = 15$ ). These specialists comprised neuro-psychologists ( $n = 2$ ), neuro-psychiatrists ( $n = 2$ ), neuro-radiologists ( $n = 4$ ), neurologists ( $n = 6$ ) and neuro-paediatrician ( $n = 1$ ). In addition to professional input, parents/carers were frequently involved in the assessment process by reporting the child's visual difficulties through structured history-taking or questionnaire completion ( $n = 18$ ). Educator input was reported in three articles. Eleven articles (23.9%) reported input from only one discipline. These included vision ( $n = 7$ ), medical ( $n = 3$ ) and psychology ( $n = 1$ ) using the groupings described previously.

### Discussion

The prevalence and awareness of CVI is increasing and, as such, there is an increased need to develop tools to aid the evaluation and diagnosis of this condition in children. This review aimed to establish what methods of assessment are currently used to investigate and diagnose CVI in children.

The overarching finding arising from a systematic review of the literature is the current lack of a standard protocol or diagnostic criteria for assessing and diagnosing childhood CVI. This review highlights that the most commonly documented presentation used to form a CVI diagnosis is based on a child presenting with visual difficulties, which are unexplained by results of vision assessment/ophthalmological examination. This 'diagnosis of exclusion' ensures that children receive a thorough examination of visual function and provides an overall profile of the child's visual function, facilitating management of co-existing ocular deficits in addition to addressing visual processing difficulties. This approach is beneficial in that it utilises equipment and assessments readily available within an ophthalmological

clinical or hospital setting, making assessment of CVI accessible and easily implemented. However, a downside to this 'diagnosis of exclusion' approach is that it may allow for diagnostic overshadowing; visual deficits recorded may be attributed to co-existing neurological impairments affecting speech, behaviour, cognition or movement rather than CVI.

Neuroimaging was the second most commonly reported method used to diagnose CVI in children. In most articles ( $n = 10$ ) this was in conjunction with visual difficulties which could not be explained by the results of vision assessment/ophthalmological examination. Often, neuroimaging was not carried out contemporaneously, but drawn from clinical records. A drawback in using this approach is the varied and often lengthy time interval between the neuroimaging assessment and subsequent clinical testing, making associations between clinical test outcomes and neuroimaging results difficult to interpret.<sup>101</sup> Lowery *et al.*<sup>102</sup> report that it is critical to have a high quality MRI scan and careful medical history to establish a diagnosis of CVI. This claim is in conflict with other authors' findings. Ortibus *et al.*<sup>47</sup> found that 14% of children with CVI in their study population presented with a normal MRI. Similarly, Franki *et al.*<sup>64</sup> sought to compare the extent and location of brain lesions using structural MRI (sMRI) in children with and without a diagnosis of CVI. They concluded that sMRI was not effective at differentiating between these groups and reported normal MRI findings in 17.4% of the population of children with CVI. Whiting *et al.*<sup>46</sup> state that while neuroimaging was useful in determining the extent and cause of brain damage in their study population, neuroimaging results did not correlate well with the degree of visual loss in patients with CVI. This contrasts with findings from Lanzi *et al.*,<sup>33</sup> who report that lesions present on MRI scans affecting the visual pathway in their study population of children with periventricular leukomalacia correlated well with visual function. In Lanzi's study cohort, children with severe damage to the optic radiations, as determined by neuroimaging, were three times more likely to have CVI compared to children with less neurological damage.<sup>33</sup> The different findings may be attributed to the variety of neuroimaging techniques employed across articles, and also how the samples have been selected in terms of CVI diagnosis. In their study, Cioni *et al.*<sup>51</sup> suggest that MRI scans play an important role in the early detection of visual deficits affecting the visual cortex and optic radiations in neonates with encephalopathy, as otherwise these deficits may continue unrecognised until children are old enough to undertake more subjective clinical measures.

Nine articles (20%) reported using structured clinical history-taking when forming a diagnosis of CVI. Applying structured history-taking tools has value in identifying key

problems with which children may present. Results from these tools can be used to develop habilitation plans for children which address highlighted problems and implement simple and practical management strategies to alleviate the impact of identified problems in daily life.<sup>8</sup> Duke *et al.*<sup>42</sup> have designed a randomised controlled trial, which is currently underway, to determine the impact of such strategies on quality of life in children with cerebral palsy and CVI. However, van Genderen *et al.*<sup>28</sup> urge caution in the use of CVI questionnaires/inventories as a screening tool (especially in isolation), and contend these tools create an unacceptable number of false positives. As such, results from structured history-taking and CVI inventories can be augmented by clinical examination or direct observation of the child.<sup>50</sup> Roman-Lantzy recommends that a child's behaviour is observed in a variety of environments and conditions, including during quiet and noisy times, with moving and stationary objects and in cluttered and uncluttered environments.<sup>70</sup> Coupling this approach with parental input can help gather a comprehensive overview of the child's visual strengths and weaknesses, and highlight areas which require habilitation. Another drawback in the use of currently available inventories to elicit difficulties associated with CVI is that they are likely to be inappropriate for infants and young children as many items rely on the child having met developmental milestones, for example walking and grasping. This is also problematic where children have co-morbidities which seriously restrict their physical or cognitive ability and for whom many of the questions applied by the inventories are not appropriate. As such, if relying solely on inventories for a diagnosis, very young or very physically or cognitively impaired children with CVI may remain undiagnosed.

Van Genderen *et al.*<sup>28</sup> sought to determine which commonly available investigative tools were effective at identifying children with CVI in a population of children with good visual acuity in a general ophthalmic clinic. They concluded that known causes of CVI in the child's medical history was the most important consideration. To further support a diagnosis, they proposed incorporation of whether the child presented with additional symptoms of cerebral damage, for example, visual field defects, nystagmus and partial optic atrophy. Consensus for this approach is evident from the present review, in which twelve articles (26.7%) considered the child's medical history to form a diagnosis in combination with other assessments.

Results of visual perception testing were reported as diagnostic in six articles (13.3%). Individual tests of visual perception often examine very specific aspects of visual processing. Due to the heterogeneous nature of CVI, if visual perception tests were used in isolation, they risk underdiagnosing children with CVI if the particular aspect of visual perception assessed is not defective. For example,



the LEA mailbox task assesses visually guided motion and perception of line direction. Even if a child performs this task without difficulty, they may exhibit other deficits in visual perception, which are not assessed using the LEA mailbox. Macintyre-Beon *et al.*<sup>37</sup> report that results on tests of visual perception do not correlate well with problems identified using the visual skills inventory questionnaire and propose that this is not a failure of the inventory, but rather the tests of visual perception as they are not developed to specifically identify problems associated with CVI. The recently developed CVIT 3-6 offers a promising alternative to previously available tests of visual perception. This test was designed specifically to identify problems associated with CVI and has shown encouraging results in stratifying children with and without a diagnosis of CVI.<sup>56</sup>

Many articles (46.7%) employed a combination of assessment methods to form a diagnosis of CVI. This approach considers multiple aspects of a child's visual function and provides a comprehensive picture of the child's visual profile. Including a range of assessments also allows flexibility in which tests are applied to each child. Affected children are likely to present with co-existing physical and mental impairments and some children may be unable to perform all tests required of them in the clinic. Implementation of a multi-assessment approach increases the likelihood of a child being able to complete some aspects of the assessment process, and therefore still provides the clinician with valuable information regarding a child's visual processing ability. While a multi-assessment approach is beneficial, it may be important to prioritise those tests which are most useful when forming a diagnosis, rather than expecting a child to complete every possible assessment method discussed in this review (and beyond).

Given that a multi-assessment approach is beneficial and often employed when diagnosing children with CVI, it is not surprising that a team of professionals may be required to apply testing. Using a team draws on multi-disciplinary expertise when assessing and interpreting results in order to ensure a consensus when providing a diagnosis.<sup>15,49</sup> In the present review, eye care professionals (specifically ophthalmologists) most commonly led the assessment process; however, this was often coupled with input from other medical and allied health professionals.

It is likely that the lack of a 'gold-standard' approach to assessing and diagnosing CVI is due to the heterogenous nature of the condition, and therefore a 'one size fits all' approach is not appropriate. Despite this, while it is accepted that a strict diagnostic framework is not a likely solution to this problem, development of 'clinical assessment guidelines' which are adaptable and inclusive to the needs of the child is warranted to ensure children receive a timely diagnosis and the support they require. It is

important that a timely CVI diagnosis can be applied, as evidence has shown that early intervention in these children improves their visual, social and educational outcomes.<sup>12,13</sup> Following diagnosis, necessary support and habilitation can be implemented in the child's home and educational environment to minimise the compounding impact CVI can have on the child's daily living. To provide an early diagnosis, it is important that assessments and tests that are developed and utilised are applicable to young paediatric patients. In addition, Philip<sup>49</sup> reports the importance of providing an assessment report to teachers and therapists involved in the child's care so that they can take account of the child's processing difficulties and ensure optimal access to education. The importance of communicating results from assessments is also echoed by Lehman<sup>103</sup> and Hyvärinen *et al.*<sup>104</sup> who recommend documenting accommodations that are medically necessary so that carers and educators can make suitable adjustments to the child's activities of daily living.

## Conclusion

The primary aim of this review was to identify and evaluate which assessments are currently used to investigate and diagnose CVI in children. Results reveal a lack of common practice in the assessment(s) utilised. A multi-assessment approach is often employed. Given the heterogeneous nature of CVI, this may be the most suitable approach with which to identify and describe CVI in a clinical situation. This approach is also beneficial in that even if a child is unable to comply with one assessment method, they may be able to comply with another to provide meaningful information on the child's CVI status. Development of sector-agreed guidelines for the assessment and diagnosis of CVI may be considered an appropriate next step in an attempt to create some clarity on when to diagnose CVI. This will ensure children receive a timely diagnosis and ultimately receive the additional support they require. However, the challenge in creating such guidelines is acknowledged due to the heterogeneity of affected children.

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## Author contributions

**Emma L McConnell:** Conceptualization (lead); data curation (equal); formal analysis (lead); investigation (lead); methodology (equal); project administration (lead); writing – original draft (lead); writing – review and editing (equal).

**Kathryn J Saunders:** Conceptualization (equal); funding acquisition (lead); supervision (equal); writing – review and editing (equal). **Julie-Anne Little:** Conceptualization (equal); data curation (equal); methodology (equal); supervision (lead); writing – review and editing (lead).

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## Supporting Information

Additional Supporting Information may be found in the online version of this article:

**Appendix S1.** Medline Search terms.

**Appendix S2.** Assessment Utility scores assigned, by category of assessment, to articles.

**Appendix S3.** List of disciplines and individuals involved in the assessment and diagnosis of cerebral visual impairment. X = input reported by article, - = input not reported by article.



**Dr Emma L McConnell** is a research optometrist at Ulster University, Queen's University Belfast and the Belfast Health and Social Care Trust. She has recently completed a PhD which investigated whether the provision of in-school vision care had measurable benefits for children attending special schools and whether investigation of cerebral visual impairment was possible as part of such a service. She is currently working on clinical trials to investigate the effect of low dose atropine eye drops on the progression of myopia in children living in the UK. Emma was recently awarded the Philip Cole Prize for practice-based research by the UK College of Optometrists.



**Professor Kathryn J Saunders** is a senior academic at Ulster University with a special interest in children's eye care. She has published over 80 peer scientific papers whose topics center on the development of visual function both in the typically developing visual system and in the presence of neurological impairment. Kathryn has led several long-term studies of typical and atypical visual development in children with, and without, learning disability. In addition to academic and research work, Kathryn delivers a teaching clinic at Ulster University where she regularly provides eye care for pre-school children and those with special educational needs. Kathryn is a Fellow of the UK College of Optometrists and is providing expert input into the development of enhanced services for children with special educational needs for NHS England.



**Julie-Anne Little** is a researcher and Associate Professor (Senior Lecturer) in the Optometry & Vision Science department within the School of Biomedical Sciences at Ulster University. Her PhD in vision science investigated the aetiology of reduced visual acuity in Down syndrome, using bespoke psychophysical techniques to parse the impact of neural, ocular and retinal deficits on visual function. Post-doctoral work in the Royal Victoria Hospital in Belfast extended her expertise in other populations with intellectual disability, including cerebral palsy and global developmental delay. In her current role as Associate Professor in Optometry at Ulster University she leads the Centre for Optometry and Vision Science. Her research continues to have a strong clinical focus, concerning the investigation of structural and functional aspects of vision, with the aim of improving vision, education outcomes and quality of life for individuals with Down syndrome and others with special needs. Publications in high-impact journals and international presentations disseminate this work, and she was returned in the UK Research Excellence Framework (REF) 2014, with the Biomedical Sciences Research Institute ranked among the top five Universities in the UK for biomedical research, and a 100% world-learning research environment. She is the Associate Research Director for Biomedical Sciences, and Research group leader for the Centre of Optometry and Vision Science. She is programme director for the MSc in Clinical Visual Science as well as a module leader for several modules in the BSc(Hons) and MOptom undergraduate Optometry programme at Ulster. She has a strong external profile in optometry as past President of the European Council of Optometry & Optics, and nationally as a Deputy Chairman of the Association of Optometrists and member of the General Optical Council's (GOC) Educational visitor panel.