Is there consensus in defining childhood cerebral visual impairment? A systematic review of terminology and definitions - Supplementary data file 1

Online literature search strategy

1. Free text search of all terms relating to CVI and childhood
2. Subject heading search of all terms relating to CVI and childhood
3. Excluding letters, editorials, comments and reviews
4. Limited to humans, English language and childhood

CVI-related free-text search terms

1. Cortical$ visual$ impair$
2. Cerebral$ visual$ impair$
3. Central$ visual$ impair$
4. Neurological$ visual$ impair$
5. Occipital$ visual$ impair$
6. Cortical$ blind$
7. Cerebral$ blind$
8. Central$ blind$
9. Neurological$ blind$
10. Occipital$ blind$

Childhood-related free-text search terms

11. Child$
12. Adolesc$
13. Infan$
14. Youth
15. Teen
16. P*ediatric
17. Baby

**possible additional letter in use*
$truncated word stems

AMED search

1. ("cerebral$ visual$ impair$" or "cortical$ visual$ impair$" or "cerebral$ blind$" or "cortical$ blind$" or "central$ visual$ impair$" or "neurological$ visual$ impair$").mp. [mp=abstract, heading words, title]
2. Child/
3. Adolescent/
4. Infant/ or Child preschool/
5. ("child$" or "adolesc$" or "teen$" or "p*ediatric" or "youth" or "infant" or "baby").mp. [mp=abstract, heading words, title]
6. 2 or 3 or 4 or 5
7. 1 and 6
8. limit 7 to (commentary or editorial or letter or "review")
9. 7 not 8

CINAHL search

#	Query	Limiters/Expanders	Last Run Via	Results
S8	S5 AND S7	Limiters - Age Groups: All Infant, All Child; Language: English Search modes - Boolean/Phrase	Interface - EBSCOhost Research Databases Search Screen - Advanced Search Database - CINAHL Plus	82
S7	S1 OR S4	Search modes - Boolean/Phrase	Interface - EBSCOhost Research Databases Search Screen - Advanced Search Database - CINAHL Plus	184
S5	("child*" or "adolesc*" or "p#ediatric" or "youth" or "infan*" or "teen" or "baby") OR MH child OR MH infant OR MH adolescence	Search modes - Boolean/Phrase	Interface - EBSCOhost Research Databases Search Screen - Advanced Search Database - CINAHL Plus	852,961
S4	MH blindness, cortical	Search modes - Boolean/Phrase	Interface - EBSCOhost Research Databases Search Screen - Advanced Search Database - CINAHL Plus	52
S1	"cortical* visual* impair*" or "cerebral* visual* impair*" or "central* visual* impair*" or "neurological* visual* impair*" or "occipital* visual* impair*" or "cortical* blind*" or "cerebral* blind*" or "neurological* blind*" or "central* blind*" or "occipital* blind*"	Search modes - Boolean/Phrase	Interface - EBSCOhost Research Databases Search Screen - Advanced Search Database - CINAHL Plus	174

EMBASE search

1. ("cerebral$ visual$ impair$" or "cortical$ visual$ impair$" or "cerebral$ blind$" or "cortical$ blind$" or "central$ visual$ impair$" or "neurological$ visual$ impair$" or "occipital$ blind$" or "central$ blind$" or "occipital$ visual$ impair$").mp. [mp=title, abstract, heading word, drug trade name, original title, device manufacturer, drug manufacturer, device trade name, keyword]
2. ("child$" or "adolesc$" or "teen$" or "p*ediatric" or "youth" or "infant" or "baby").mp. [mp=title, abstract, heading word, drug trade name, original title, device manufacturer, drug manufacturer, device trade name, keyword]
3. cerebral blindness/
4. childhood/
5. infancy/
6. adolescence/
7. 1 or 3
8. 2 or 4 or 5 or 6
9. 7 and 8
10. limit 9 to child
11. limit 10 to human
12. limit 11 to (editorial or letter or "review")
13. 11 not 12

MEDLINE search

1. ("cerebral$ visual$ impair$" or "cortical$ visual$ impair$" or "cerebral$ blind$" or "cortical$ blind$" or "central$ visual$ impair$" or "neurological$ visual$ impair$" or "occipital$ blind$" or "central$ blind$" or "occipital$ visual$ impair$").mp. [mp=title, abstract, original title, name of substance word, subject heading word, keyword heading word, protocol supplementary concept word, rare disease supplementary concept word, unique identifier]
2. Blindness, Cortical/
3. ("child$" or "adolesc$" or "teen$" or "p*ediatric" or "youth" or "infant" or "baby").mp. [mp=title, abstract, original title, name of substance word, subject heading word, keyword heading word, protocol supplementary concept word, rare disease supplementary concept word, unique identifier]
4. Child/
5. Adolescent/
6. Infant/
7. 1 or 2
8. 3 or 4 or 5 or 6
9. 7 and 8
10. limit 9 to humans
11. limit 10 to "all child (0 to 18 years)"
12. limit 11 to (comment or editorial or letter or "review")
13. 11 not 12

PSYCHINFO search

1. ("cerebral$ visual$ impair$" or "cortical$ visual$ impair$" or "cerebral$ blind$" or "cortical$ blind$" or "central$ visual$ impair$" or "neurological$ visual$ impair$" or "occipital$ blind$" or "central$ blind$" or "occipital$ visual$ impair$").mp. [mp=title, abstract, heading word, table of contents, key concepts, original title, tests & measures]
2. ("child$" or "adolesc$" or "teen$" or "p*ediatric" or "youth" or "infant" or "baby").mp. [mp=title, abstract, heading word, table of contents, key concepts, original title, tests & measures]
3. 1 and 2
4. limit 3 to human
5. limit 4 to (childhood or adolescence <13 to 17 years>)
6. limit 5 to ("comment/reply" or editorial or letter or reviews)
7. 5 not 6

Table S1: Online Search Subject Headings and Limits

Database	Vision	Age	Limits	Search 03.01.2016
AMED	N/A	Child Infant Adolescent Child, preschool	N/A	4 results
CINAHIL	Blindness, cortical	childhood	All child All infant	82 results
Embase	Cerebral blindness	Childhood Infancy adolescence	Human Child unspecified age	441 results
Medline	Blindness, cortical	Child Infant Adolescent	Humans All child	561 results
PsychInfo	N/A	N/A	Human Childhood Adolescence	90 results

Manual search of textbooks:

Roman-Lantzy, C. (2007). *Cortical visual impairment: An approach to assessment and intervention*. AFB Press.

Dutton, G., & Bax, M. (2010). *Visual impairment in children due to damage to the brain: Clinics in Developmental Medicine* (Vol. 10). John Wiley & Sons.

Zihl, J., & Dutton, G. N. (2015). *Cerebral Visual Impairment in Children*. Springer Vienna.

Lueck, A. H. & Dutton, G. N. (2015). *Vision and the Brain: Understanding Cerebral Visual Impairment in Children*. AFB Press.

CVI SYSTEMATIC REVIEW DATA EXTRACTION FORM

Study ID	Reviewer, Date	Checked by

ARTICLE DEMOGRAPHICS	
Author, Year	
Journal	
Country of origin (research group)	
CVI INFORMATION	
Full CVI term used	
Definition of CVI (if applicable)	Page:_____
Diagnostic criteria of CVI (if applicable)	Page:_____
Assessment tools used for CVI identification	
SAMPLE CHARACTERISTICS	
Sample studied and restrictions? (i.e. only subset of children with CVI included) **Why are there restrictions?**	
Extent of disease **(range of CVI symptoms)**	
Additional diagnoses in group	
Total and CVI sample size	
Total and CVI sample age range	
Total and CVI sample gender distribution	
Main findings of the study	

Table S2: Full definitions and diagnostic criteria of included articles

Article ID	Definition	Diagnostic criteria
Frank & Torres (1979)	Cortical or cerebral blindness is defined as bilateral absence of vision caused by cerebral disease [4] (I)	All had neurological and ophthalmological examinations. None had any ocular abnormality, and all had normal pupillary responses and normal funduscopic examinations. Optokinetic nystagmus was absent in patients in whom this was examined. The patients showed no response to visual threat, did not track objects, and lacked all be- havioral visual responses (M)
Mohn *et al.* (1983)	However, there are now serious doubts as to the diagnostic value of flash-VEPs, since positive, although often abnormal flash-VEPs may be recorded in children diagnosed as cortically blind, i.e. blind in the absence of ocular abnormalities [1, 7] (I)	Two children were diagnosed as cortically blind, i.e. blind in the absence of ocular abnormalities, and with intact pupillary reactions. (R)
Robertson *et al.* (1986)	Cortical blindness traditionally has been defined as absence of all visual sensation, normal ocular examination, loss of optokinetic nystagmus, preservation of pupillary response and normal mobility of the eyes. Recent studies, however, showed that this concept of cortical blindness is too restrictive. Cortical visual loss covers a spectrum ranging from no apparent vision to residual sight and not infrequently it coexists with ocular damage due to the wide distribution of cerebral insults. Since blindness implies total absence of vision, cortical visual loss or cortical visual impairment (CVI) is a more appropriate term. (I)	CVI was suspected when the degree of visual loss was unexplained by the ocular examination alone. It was diagnosed in the presence of severe visual loss, normal or minimal ocular findings and clinical electrodiagnostic and CT evidence of postgeniculate lesions involving the visual cortex. (M)
Roland *et al.* (1986)	The conventional definition of cortical blindness implies absence of all visual sensation and opticokinetic nystagmus, with preservation of pupillary light responses and normal ocular examination [5]. However, recent data suggest a spectrum of cortical visual loss ranging from total blindness to significant residual sight [1,3,4,6]. Because blindness implies total absence of vision, cortical visual impairment (CVI) is a more accurate term [3].. (I)	CVI was suspected when the extent of visual loss was unexplained by ocular abnormalities. (M)
Jan *et al.* (1987)		CVI was suspected when the degree of visual loss was unexplained by the ocular examination alone. It was diagnosed in the presence of severe visual loss, normal or minimal ocular findings and clinical, electrodiagnostic and CT evidence of post-geniculate lesions involving the visual cortex. (M)
Bencivenga *et al.* (1989)	In this study, the relationship of the electrical response to photic stimulation at different scalp locations is examined in children with damage to the striate cortex (area 17) or to the association areas (areas 18-22). The purpose was to develop objective parameters derived from the VEP which might complement clinical localization in children with cortical visual impairment (CVI) resulting from involvement in these areas.(I)	All had severe visual loss, normal or minimal ocular findings, and clinical and computed tomography (CT) evidence of a post-geniculate abnormality. (M)
Flodmark *et al.* (1990)	CVI was defined as visual loss caused by a disturbance of the posterior visual pathway and/or visual cortex (M)	
Taylor & McCulloch (1991)	We used the definition of cortical blindness as stated by Barnet et al. "the absence of vision in the presence of normal pupillary reflexes, without significant ophthalmological disease" [11]. (M)	
Wong (1991)	Cortical blindness (CB) has been defined as complete loss of all visual sensation and loss of optokinetic nystagmus with preservation of pupillary response, normal eye motility, and normal retina [1]. Cortical visual impairment (CVI) has been recommended as a better term because it encompasses a spectrum of visual loss ranging from absent to some residual vision [2,3] (I)	Patients were included when severe visual loss occurred in the presence of normal pupillary response and normal fundi. Patients were excluded when the duration of visual loss lasted less than 48 hours. (M)
Chen *et al.* (1992)	cortical visual impairment results from bilateral postchiasmatic cerebral lesions. Infants with cortical visual impairment have poor vision, absent optokinetic nystagmus, normal pupillary reflexes, and normal ocular structure. (I)	Cortical visual impairment was diagnosed if the infant had poor vision at initial examination, absent optokinetic nystagmus, normal pupillary reflexes, and normal ocular structure. (M)
Frank *et al.* (1992)	Cerebral blindness is a clinical syndrome manifested by a complete loss of visual behavior, including appreciation of light and loss of optokinetic nystagmus. The media and retina of the eyes are normal, pupillary responses to light are intact and there is no optic atrophy. The lesion responsible is thought to affect the visual pathways beyond the lateral geniculate nucleus and/or cortical areas involved in vision. (I)	All children had normal eye examinations, and patients with any evidence of ocular, retinal or prechiasmal visual abnormalities were excluded. To confirm the clinical diagnosis of complete cerebral blindness, the visual function of each child was tested behaviorally by placing high-contrast black and white bullseye, checkerboard and striped patterns in front of the child's eyes at a distance of approximately 18cm (M)
Schenk-Rootlieb *et al.* (1992)	He defined cerebral visual disturbance (CVD) as a disturbance of vision caused by defective function of the retrochiasmatic part of the visual system, and demonstrated that CVD formed part of the CP syndrome. (I)	An ophthalmological explanation for low visual acuity could be provided for only seven of these 43 patients; therefore CVD is likely to be present in 36 of the 43 patients'(84 per cent) with low visual acuity on both methods. (D)

Article ID	Definition	Diagnostic criteria
Granet *et al.* (1993)	Poor visual function in an infant with a normal anterior visual pathway is a prognostic dilemma for an ophthalmologist. Central visual impairment is suggested as a term that describes this disorder and includes both acquired and congenital abnormalities. (I)	From this group, 70 patients had central visual impairment defined as follows: (1) clinical visual impairment, (2) an abnormal initial visual-evoked response, (3) the presence of normal ocular structures, and (4) a normal electroretinogram. (M)
Jan *et al.* (1993)	CVI was defined as a disturbance of the posterior visual pathways and/or occipital lobes leading to loss of sight. (M)	
Schenk-Rootlieb *et al.* (1993)	Cerebral visual impairment, defined by Van Nieuwenhuizen (1987) as defective visual function caused by a lesion of the retrochiasmatic part of the central visual system, (I)	In the remaining 43 patients no ophthalmological explanation could be found for the low acuity and, therefore, there was a probability of cerebral visual impairment. (Patients section)
Schenk-Rootlieb *et al.* (1994)	In addition to ophthalmological disorders, such as squint and refraction anomalies, CVI, defined by Van Nieuwenhuizen (17) as a disturbance of vision caused by defective function of the retrochiasmatic part of the visual system, is frequently present (I)	The subnormal acuity found in 50 patients could be explained ophthalmologieally in only 7. In the remaining 43 patients, therefore, the visual impairment must be due to cerebral involvement, as deseribed earlier (13) (M)
Eken *et al.* (1995)		It was decided that repeated acuity estimates at or below the 10th centile would be considered as indicative of cerebral visual impairment, when the extent of visual loss was unexplained by ocular abnormalities (M)
Cioni *et al.* (1996)	Many authors think that 'cerebral visual impairment' (CVI) is a better term than 'cortical blindness' (Hertz et al. 1988; Eken et al. 1994, 1995; Schenk-Rootlieb et al. 1994) to describe poor visual function in children caused by abnormalities of the posterior visual pathway Lesions inducing a visual deficit may involve different pathways and cortical areas of the developing brain and not necessarily the primary visual cortex only. Moreover, since the majority of subjects with CVI have some residual vision, visual impairment seems to be a more appropriate term than blindness. (I)	CVI was found in 48 patients (Table II). 10 of them were considered totally blind because they did not show any response to any of the stimuli: acuity cards, mother's face or objects. The other 38 children with CVI showed a severe loss of vision, i.e. acuity values below the 5th centile, according to the ACP. (R)
Eken *et al.* (1996)		CVI was diagnosed in the presence of severe visual loss, when the extent of visual impairment could not be explained by ocular abnormalities. "Severe visual loss" was considered when an infant had acuity card estimates at or below the 10th centile on at least two consecutive occasions including the test at 18 months. (M)
Kwok *et al.* (1996)		if the result of the acuity measurement was poor despite normal ocular structures and pupillary responses, these patients were regarded to have cortical visual impairment (CVI) (Whiting et al 1985) (M)
Uggetti *et al.* (1996)	Cerebral visual impairment was defined as reduced sight despite normal ophtalmologic findings or findings too mild to explain the visual loss (20). (M)	
Cioni *et al.* (1997)	Visual disorders of preterm infants include abnormalities of the anterior visual pathways (eye to optic chiasm), mainly retinopathy of prematurity, but also cerebral visual impairment (CVI), i.e., a visual loss due to a disturbance of posterior visual pathways. (I)	
Lanzi *et al.* (1998)	CVI has been defined as a reduced visual function, in the absence of sufficient ophthalmological data to explain the deficit (20). (M)	
Stiers *et al.* (1998)	Cerebral visual impairment (CVI) in chlldren is a condition of reduced visual capacity resulting from damage to the visual pathways and projection areas posterior to the optic chiasm (11, 29, 30, 35). (I)	They were included because of indications of visual perceptual problems, in the absence of a satisfactory ophthalmologic explanation, thus making CVI highly probable. (M)
Huo *et al.* (1999)		At the time of the patient's appointment, the diagnosis of "CVI" was based upon (1) vision loss in the absence of signs of anterior visual pathway disease, or (2) vision loss greatly exceeding that which would be expected, given the findings of an ocular examination. (M)
Oud *et al.* (1999)	In the present study, CVI is defined as a disturbance of vision caused by the defective function of the retrochiasmatic part of the visual system.14 (I)	If the child had normal ocular structures, normal pupil reactions to light, and a normal fundus without severe pallor of the optic disc, the problem was considered to be located in the retrochiasmatic pathways and, according to our definition, the child was diagnosed as suffering from CVI. (D)
Choi *et al.* (2001)	Cortical visual impairment (CVI) refers to the loss of vision accompanying injuries to, or maldevelopment of, the geniculate and/or extrageniculate pathways.1 (I)	It involved children with CVI, who had impairment of vision with normal pupillary response and no other abnormalities in either the anterior or the posterior segment, including the optic disc (M)
Good (2001)	Cortical visual impairment is a neurologic impairment defined as bilateral loss of central vision (visual acuity) caused by damage to the central nervous system. In other words, visual acuity is reduced as a result of nonocular disease (3,4). (I)	CVI was diagnosed on the basis of poor visual attention or behavior associated with a normal ophthalmologic exami- nation and normal pupillary responses (M)

Article ID	Definition	Diagnostic criteria
Weiss *et al.* (2001)	In this study, CVI was defined as bilateral visual dysfunction caused by cortical disturbances in patients with a normal eye examination and normal anterior visual path- ways. We use the term visual dysfunction to emphasize that these patients have more than just acuity deficits (D)	The study population consisted of 31 of 63 consecutive infants who were visually unresponsive despite having clear ocular media and normal fundi (M)
Brodsky *et al.* (2002)	the umbrella term cortical visual loss currently comprises two subgroups: one having term injury that predominantly involves the striate and peristriate cortex and the other having preterm injury that predominantly involves the subcortical white matter, including the optic radiations. (I)	Retrogeniculate visual loss was diagnosed clinically when bilateral visual impairment was accompanied by other neurologic dysfunction and when the degree of visual impairment was out of proportion to any ocular abnormalities (M)
Sakai *et al.* (2002)	Cerebral visual impairment is defined as reduced visual capacity resulting from damages in the retrochiasmatic visual pathways and areas.1–3 (I)	Diagnosis of cerebral visual impairment was made when the following five conditions were satisfied: (1) presence of normal pupillary responses,13 (2) absence of ocular diseases,14 (3) presence of damage in the posterior visual pathway or occipital cortices as suggested by neurologic findings15 and history, (4) extreme restriction of visual behavior,14 and (5) presence of persistent impairment of binocular vision.13 (M)
Hoyt (2003)		At the time of the child's initial appointment, a diagnosis of cortical visual impairment was made if there was (1) visual loss in the absence of signs of anterior visual pathway disease or (2) vision loss exceeding that, which was expected given the findings of the ocular examination. (M)
Skozenski & Good (2004)	It is a condition in which children have reduced visual acuity as a result of damage to posterior visual pathways (Good et al. 1994, 2001b) (I)	CVI was diagnosed when vision was clearly subnormal from a clinical standpoint, in the context of a normal, or nearly normal eye examination. (M)
Sie *et al.* (2005)	Cerebral visual impairment is often present as a result of damage to the retrochiasmatic pathways and is particularly common in cases in which hypoxic-ischemic damage involves the periventricular occipital white matter, as shown by late MRI [11,15,23, 25,28,38,42] (D)	In the absence of ophthalmological abnormalities, the decreased vision was ascribed to cerebral visual impairment. (M)
Good & Hou (2006)	The leading cause of vision impairment in children in the western world is cortical or cerebral visual impairment (CVI), caused by damage to the visual cortex or optic radiations.1,2 (I)	CVI diagnosed clinically on the basis of reduced visual acuity with preserved pupillary reactions and normal results in eye examinations (M)
Matsuba & Jan (2006)	As CVI results from damage to the posterior visual pathways, the increase in the reported incidence is likely the result of increased recognition as well as advances in medical care (Good et al. 2001) (I)	clinical characteristics consistent with CVI, including but not limited to poor visual attention, light gazing, and photophobia (M)
Fazzi *et al.* (2007)	Cerebral visual impairment is defined as a deficit of visual function caused by damage to, or malfunctioning of, the retrogeniculate visual pathways (optic radiations, occipital cortex, associative visual areas) in the absence of any major ocular disease.2. (I)	This study sample included all those with central nervous system abnormalities who, at the end of the evaluation detailed above, were deemed to be affected by cerebral visual impairment, diagnosed according to the criteria set out in the introduction.2,5 (M)
Khetpal & Donahue (2007)	CVI is commonly defined as a loss in visual function in the absence of damage to the anterior afferent visual pathways or ocular structures. CVI can be defined as a decrease in visual acuity or visual fields of a person with structurally normal eyes and normal-appearing anterior afferent visual pathways (I)	The diagnosis of CVI was based on an ocular examination that revealed poor visual function bilaterally that could not be accounted for by age, structural ocular examination findings, optic atrophy, or high refractive error. (M)
Ghasia *et al.* (2008)	For the present study, CVI was defined as bilateral, subnormal, best corrected visual acuity for age that could not be attributed to an ocular motor deficit (e.g., nystagmus) or a structural defect of the anterior afferent visual pathway (e.g., bilateral optic neuropathy)—the standard definition of CVI used in North America.44–50 (M)	
Ferziger *et al.* (2011)	CVI is defined as a bilateral loss of central visual function (visual acuity) caused by neurological damage to the visual cortex and⁄or visual pathway structures. (I)	A diagnosis of CVI was made if there was poor bilateral visual function despite a normal anterior pathway eye examination or which could not be accounted for based on the clinical examination. 6,7,16 (M)
Good *et al.* (2012)		Children with CVI were diagnosed clinically on the basis of reduced visual acuity in both eyes, with the diagnosis corroborated by neuroimaging, history, and physical examination to exclude with certainty any coexisting eye disease. Pupillary reactions were normal. (M)
Weinstein *et al.* (2012)	CVI may include damage to one or more visual structures including the optic radiations, primary visual cortex, association visual cortices, and white matter pathways that connect visual cortical areas. Damage to primary visual cortex or precortical pathways produces visual field defects1,2 that may contribute to impairment of both primary and secondary visual process- ing.1,2,4 (I)	CVI was diagnosed when visual acuity was clearly outside the typical range for age in both eyes,15,16 was not explained by ocular abnormalities, and was accompanied by structural changes of either PVL or hydrocephalus involving the parietal and⁄or occipital lobes on either magnetic resonance imaging (18 individuals) or ultrasound (one individual with PVL). Although the spectrum of CVI properly includes some individuals with normal or near-normal visual acuity but with deficits of higher order visual function, we did not include children with normal acuity in the study. Although exclusion of these individuals may create some selection bias, we chose visual acuity loss as an easily defined and readily applicable operational criterion for entry into the study, although it is not a defining feature of CVI. (M)

Article ID	Definition	Diagnostic criteria
Bosch *et al.* (2014a)	In the absence of absolute criteria for the diagnosis, the following definition is commonly used: CVI includes all visual dysfunctions caused by damage to, or malfunctioning of, the retrochiasmatic pathways in the absence of any major ocular disease [3]. A more practical definition includes an impairment of vision with normal function of the ocular structures and anterior visual pathways [4-6)(I)	CVI was diagnosed when there was no other ocular diagnosis which could explain the visual impairment or visual field defect, and/or typical features such as poor fixation or crowding, and/or CVI were found at neuropsychological investigation. Neuropsychological investigation of the visual functions was, however, not possible in a majority of the individuals, because of their developmental age. (M)
Bosch *et al.* *(*2014b)	It includes all visual dysfunctions caused by damage to, or malfunctioning of, the retrochiasmatic pathways in the absence of damage to the anterior visual pathways or any major ocular disease.3,4 (I)	CVI was diagnosed by a pediatric ophthalmologist under the following following criteria: no other ocular diagnosis which could explain the visual impairment or visual field defect, and/or typical features such as poor fixation or crowding, and/or CVI found at neuropsychological investigation. Neuropsychological investigation was, however, not possible in a majority of the individuals, because of their developmental age. (M)
Cavascan *et al.* (2014)	Cerebral visual impairment (CVI) is a pediatric neurological disorder caused by post-chiasmatic brain lesion that results in bilateral visual loss [1, 2]. Clinically, it manifests as a deficit in visual acuity with normal ocular structures in both eyes [1, 2]. (I)	Children with CVI were diagnosed clinically on the basis of bilaterally reduced visual acuity, with evidence of post-chiasmatic brain lesion, poor visual behavior and normal ocular structure. (M)
Chong & Dai (2014)	The major causes of visual impairment differ widely by region,3 but in developed nations the leading cause of childhood visual impairment is cerebral visual impairment (CVI),4 which is thought to result from an increased survival rate of premature infants and neonates with complex medical requirements.5,6 CVI is also known as cortical visual impairment and cortical blindness. The term cerebral visual impairment is used in this study because the cortex is rarely affected in isolation.6 CVI is caused by pathology to the retrochiasmatic pathway. Hence CVI should not be diagnosed if there is concurrent pathology of the anterior visual pathway.7 CVI is caused by pathology to the retrochiasmatic pathway. Hence CVI should not be diagnosed if there is concurrent pathology of the anterior visual pathway.7 (I)	Inclusion criteria for this study included BLENNZ registered children, aged #16 years of age with a confirmed diagnosis of CVI and visual acuity of #6/18 in the better-seeing eye….In this study, the diagnosis of CVI was made only when there was no coexisting anterior visual pathway pathology or refractive error. (M)
Geldof *et al.* (2015)	Our functional approach to defining CVI included any oculomotor, visual sensory, and perceptive deficit arising from cerebral abnormalities, thereby extending the range of deficits in conventional definitions of CVI (3,5,6). Our approach incorporates recent insights indicating that also “sensory” functions of binocular vision such as stereopsis have their cerebral underpinnings (24). Consequently, CVI becomes a broad umbrella term that includes a wide range of visual deficits. (D)	The diagnosis of CVI was determined based on the results of ophthalmological and psychological /neuropsychological research and on the assessment data reported by a developmental coach specialised in working with children with visual impairments, using the following criteria: a normal or near-normal eye exam (corrected vision >0.3 and/or field of vision >308) performed by an ophthalmologist; a history or presence of neurological problems; and presence of behavioural responses to visual stimuli that are unique to CVI. This results in strong colour preferences, need for movement to elicit or sustain visual attention, visual latency-delayed responses in looking at objects, visual field preferences, difficulties with visual complexity, light-gazing and non-purposeful gaze, difficulty with distance viewing, and absent or atypical visual reflexes (Dutton & Jacobson, 2001; Stiers et al., 2002). (M)
Mezer *et al.* (2015)		CVI was usually diagnosed by an ophthalmologist as bilaterally diminished visual acuity caused by damage shown on MRI to the occipital lobes or to the visual pathway (M)
Binder *et al.* (2016)	CVI is a clinical diagnosis, in which there is no ocular or anterior visual pathway pathology sufficient to explain the level of impairment. (I)	CVI was diagnosed clinically as visual impairment without ocular or anterior visual pathway pathology sufficient to explain the level of impairment (M)
Kemmanu *et al.* (2016)		Children who had sub-normal vision with a normal retina on indirect ophthalmoscopy and had no evidence of nystagmus were diagnosed as having cortical visual impairment. (M)
Özturk *et al.* (2016)		Bilateral decreased visual response caused by damage to either the visual cortex or the geniculostriate visual pathways based on MRI findings was defined as cortical visual impairment (CVI), unless any ocular abnormalities related to visual loss existed. (M)

I: Introduction
M: Methods
R: results
D: Discussion

Table S3: methodological details of included studies

Article ID	Condition studied	Control/ other group?	Study design	CVI diagnosed in study or before	Total sample size	CVI sample size	CVI sample age range (years)	CVI sample percentage male	Need for VI in the CVI group*	Co-occurring ocular pathology excluded?
Frank & Torres (1979)	CVI	yes	cross-sectional	prior to study	61	30	0-15	NR	yes	excluded
Mohn *et al.* (1983)	CVI	yes	cross-sectional	prior to study	37	2	NR	NR	yes	excluded
Robertson *et al.* (1986)	CVI	no	cross-sectional	prior to study	40	40	1-15	NR	yes	excluded
Roland *et al.* (1986)	CVI and hypoxia/ischaemia	no	longitudinal	prior to study	20	20	0-17	NR	yes	included
Jan *et al.* (1987)	CVI	no	cross-sectional	prior to study	50	50	0-17	NR	yes	included
Bencivenga *et al.* (1989)	CVI	yes	cross-sectional	prior to study	78	26	1-13	NR	yes	excluded
Flodmark *et al.* (1990)	CVI	no	cross-sectional	prior to study	95	95	NR	NR	yes	excluded
Taylor & McCulloch (1991)	CVI	no	longitudinal	prior to study	32	32	0-5	NR	yes	excluded
Wong (1991)	CVI	no	cross-sectional	prior to study	32	32	0-14	59	yes	excluded
Chen *et al.* (1992)	CVI	no	longitudinal	prior to study	30	30	1	43	yes	excluded
Frank *et al.* (1992)	CVI	yes	cross-sectional	prior to study	120	60	0-10	NR	yes	excluded
Schenk-Rootlieb *et al.* (1992)	CP	yes	longitudinal	during study	164	43	NR	NR	yes	included
Granet *et al.* (1993)	CVI	no	longitudinal	prior to study	10	10	0-4	NR	yes	excluded
Jan *et al.* (1993)	CVI and another factor	no	cross-sectional	prior to study	83	83	NR	61	yes	excluded
Schenk-Rootlieb *et al.* (1993)	CP	yes	cross-sectional	during study	67	43	NR	NR	yes	excluded
Schenk-Rootlieb *et al.* (1994)	CP	yes	cross-sectional	during study	74	43	NR	NR	yes	included
Eken *et al.* (1995)	leucomalacia/posterior pathway damage	yes	longitudinal	during study	65	11	0-2	55	yes	included
Cioni *et al.* (1996)	leucomalacia/posterior pathway damage	yes	cross-sectional	during study	80	48	0-7	50	yes	included
Eken *et al.* (1996)	CVI and leucomalacia	no	longitudinal	during study	9	9	0-2	40	yes	included
Kwok *et al.* (1996)	other	no	cross-sectional	during study	260	31	NR	NR	yes	excluded
Uggetti *et al.* (1996)	CP	yes	cross-sectional	during study	27	17	NR	41	yes	included
Cioni *et al.* (1997)	leucomalacia/posterior pathway damage	yes	longitudinal	during study	66	37	NR	NR	yes	excluded
Lanzi *et al.* (1998)	CP	yes	cross-sectional	during study	38	23	NR	NR	yes	included
Stiers *et al.* (1998)	CVI	yes	cross-sectional	prior to study	102	22	2-16	72	no	included
Huo *et al.* (1999)	CVI	no	cross-sectional	prior to study	170	170	NR	NR	yes	included
Oud *et al.* (1999)	CVI and another factor	no	cross-sectional	during study	72	32	0-16	NR	yes	included
Choi *et al.* (2001)	CVI	yes	cross-sectional	prior to study	20	8	1-12	38	yes	excluded
Good (2001)	CVI	no	cross-sectional	prior to study	41	41	1-16	NR	no	excluded
Weiss *et al.* (2001)	CVI	yes	longitudinal	prior to study	62	31	0-5	52	yes	excluded
Brodsky *et al.* (2002)	leucomalacia/posterior pathway damage	no	cross-sectional	prior to study	100	100	NR	NR	yes	included
Sakai *et al.* (2002)	CVI and another factor	yes	cross-sectional	prior to study	21	9	4-35	78	yes	excluded
Hoyt (2003)	CVI	no	longitudinal	prior to study	67	67	NR	NR	yes	included
Skozenski & Good (2004)	CVI	no	cross-sectional	prior to study	35	35	0-16	57	yes	included
Sie *et al.* (2005)	leucomalacia/posterior pathway damage	no	longitudinal	during study	46	46	0-12	76	yes	excluded
Good & Hou (2006)	CVI	yes	cross-sectional	prior to study	37	20	1-5	NR	yes	excluded
Matsuba & Jan (2006)	CVI	no	longitudinal	prior to study	423	423	NR	53	yes	included
Fazzi *et al.* (2007)	CVI	no	cross-sectional	prior to study	121	121	0-15	41	no	included
Khetpal & Donahue (2007)	CVI	no	cross-sectional	prior to study	98	98	0-19	57	yes	included

Article ID	Condition studied	Control/ other group?	Study design	CVI diagnosed in study or before	Total sample size	CVI sample size	CVI sample age range (years)	CVI sample percentage male	Need for VI in the CVI group*	Co-occurring ocular pathology excluded?
Ghasia *et al.* (2008)	CP	yes	cross-sectional	during study	50	8	2-19	NR	yes	included
Ferziger *et al*. (2011)	CP	yes	cross-sectional	during study	77	26	3-20	50	yes	included
Good *et al.* (2012)	CVI	yes	cross-sectional	prior to study	50	34	1-6	65	yes	excluded
Weinstein *et al.* (2012)	CVI and leucomalacia	yes	cross-sectional	prior to study	100	19	NR	47	yes	excluded
Bosch, Willemse *et al.* (2014)	CVI	no	cross-sectional	prior to study	309	309	0-45	55	yes	excluded
Bosch, Boonstra *et al. (*2014)	CVI and another factor	no	cross-sectional	prior to study	197	197	NR	NR	yes	excluded
Cavascan *et al.* (2014)	CVI	no	cross-sectional	prior to study	115	115	0-13	57	yes	excluded
Chong & Dai (2014)	CVI	no	cross-sectional	during study	182	182	0-16	64	yes	excluded
Geldof *et al.* (2015)	at risk of CVI	yes	cross-sectional	during study	167	25	5	52	no	included
Mezer *et al.* (2015)	general VI	yes	cross-sectional	prior to study	NR	NR	0-18	NR	yes	NR
Binder *et al.* (2016)	CVI and another factor	no	longitudinal	prior to study	70	70	1-9	NR	yes	included
Kemmanu *et al.* (2016)	general VI	yes	cross-sectional	during study	615	34	NR	NR	yes	excluded
Özturk *et al.* (2016)	general VI	yes	cross-sectional	prior to study	695	212	NR	NR	yes	excluded

* Did the participant need to have a visual impairment (reduced visual acuity/fields) to be included in the CVI sample?
NR: Not reported

Table S4: assessment information of included studies

	Eye examination*								Co-occurring conditions in the sample					
Article ID	**Assessed by**	**fundus exam**	**refraction**	**ocular motility/alignment**	**visual deficits assessment**	**neuroimaging**	**visual electrophysiology included (at least subgroup)**	**cognitive/ neuropsychological assessment**	**intellectual disability/ developmental delay**	**movement disorder**	**seizure disorder**	**hydrocephalus/ myelomeningocele**	**Hearing impairment**	**Other conditions**
Frank & Torres (1979)	NR	+	-	-	VA assumed from inclusion criteria	-	+	-	+	+	-	-	-	CNS degenerative disease
Mohn *et al.* (1983)	NR	-	-	-	VA, VF	CT	+	-	-	-	-	-	-	
Robertson *et al.* (1986)	MDT	-	-	-	LV	CT	+	-	+	+	-	+	-	
Roland *et al.* (1986)	MDT: ophthalmologist, paediatric neurologist, psychologist, physiotherapist, speech and language therapist	-	-	-	VA, LV	CT	+	cog, VP	+	+	+	-	-	
Jan *et al.* (1987)	MDT: ophthalmologist, paediatric neurologist, psychologist, physiotherapist, speech and language therapist	-	-	-	LV, VF	CT	+	NR	+	+	+	+	-	
Bencivenga *et al.* (1989)	NR	-	-	-	VA assumed from inclusion criteria	CT	+	-	+	+	+	+	-	
Flodmark *et al.* (1990)	NR	-	-	-	LV	CT	+	NR	+	+	+	-	-	
Taylor & McCulloch (1991)	NR	-	-	-	LV	-	+	-	-	-	-	-	-	
Wong (1991)	MDT: ophthalmologist, developmental paediatrician, child neurologist	-	-	-	LV	CT	+	-	+	+	+	-	-	
Chen *et al.* (1992)	developmental assessment by paediatrician	+	+	+	LV	MRI/ CT	-	cog	+	+	+	+	+	
Frank *et al.* (1992)	NR	-	-	-	LV	-	+	-	+	-	+	-	-	
Schenk-Rootlieb *et al.* (1992)	NR	-	-	-	VA	.	-	-	-	+	-	-	-	
Granet *et al.* (1993)	Paediatric ophthalmologist	+	+	+	VA	.	+	-	-	-	-	-	-	

	Eye examination*								Co-occurring conditions in the sample					
Article ID	**Assessed by**	**fundus exam**	**refraction**	**ocular motility/alignment**	**visual deficits assessment**	**neuroimaging**	**visual electrophysiology included (at least subgroup)**	**cognitive/ neuropsychological assessment**	**intellectual disability/ developmental delay**	**movement disorder**	**seizure disorder**	**hydrocephalus/ myelomeningocele**	**Hearing impairment**	**Other conditions**
Jan *et al.* (1993)	MDT	-	-	-	VA, VF	CT	+	-	+	+	+	+	-	
Schenk-Rootlieb *et al.* (1993)	NR	-	-	-	VA	-	-	cog	+	+	-	-	-	
Schenk-Rootlieb *et al.* (1994)	NR	-	-	-	VA	CT	-	-	+	+	+	-	-	
Eken *et al.* (1995)	eye exam by ophthalmologist	-	+	-	VA	US/MRI/ CT	-	cog	+	+	+	-	-	
Cioni *et al.* (1996)	NR	+	-	+	VA, VF	MRI	+	-	+	+	-	-	-	
Eken *et al.* (1996)	eye exam by ophthalmologist	-	+	-	VA	US/MRI/ CT	+	cog	+	+	+	-	-	
Kwok *et al.* (1996)	MDT: Developmental paediatrician, audiologist, nursing, teacher, medical social worker	+	+	+	VA, LV	-	-	-	+	+	-	-	-	
Uggetti *et al.* (1996)	child neuropsychiatrist	+	-	-	VA	MRI	-	-	-	+	-	-	-	
Cioni *et al.* (1997)	NR	-	-	+	VA, VF	US/MRI	-	-	-	+	-	-	-	
Lanzi *et al.* (1998)	child neuropsychiatrist	+	+	+	VA	MRI	+	cog	-	+	-	-	-	
Stiers *et al.* (1998)	MDT	+	+	+	VA, neuropsych.	MRI/ CT subgroup	-	cog, VP	-	+	-	-	-	DCD, PDD
Huo *et al.* (1999)	NR	-	-	-	LV, VF	-	-	-	-	+	+	-	+	developmental regres
Oud *et al.* (1999)	eye exam by ophthalmologist	+	+	+	VA	CT/MRI	-	-	-	-	-	+	-	
Choi *et al.* (2001)	NR	-	-	-	VA	PET	+	-	-	-	-	-	-	
Good (2001)	NR	-	-	-	VA, LV	-	+	-	-	-	-	-	-	

	Eye examination*								Co-occurring conditions in the sample					
Article ID	**Assessed by**	**fundus exam**	**refraction**	**ocular motility/alignment**	**visual deficits assessment**	**neuroimaging**	**visual electrophysiology included (at least subgroup)**	**cognitive/ neuropsychological assessment**	**intellectual disability/ developmental delay**	**movement disorder**	**seizure disorder**	**hydrocephalus/ myelomeningocele**	**Hearing impairment**	**Other conditions**
Weiss (2001)	NR	-	-	-	VA	CT, MRI	+	-	+	+	+	-	-	
Brodsky *et al.* (2002)	NR	+	-	+	VA	MRI/CT	-	-	-	-	-	-	-	
Sakai *et al.* (2002)	NR	-	-	+	LV, CS	-	+	-	+	+	-	-	-	
Hoyt (2003)	NR	-	-	-	LV	CT/MRI	+	-	-	-	-	-	-	
Skozenski & Good (2004)	NR	-	-	-	VA	-	+	-	-	-	+	-	-	
Sie *et al.* (2005)	paediatric neurologist for VA	-	-	-	VA	MRI	-	cog	+	+	-	-	-	
Good & Hou (2006)	NR	-	-	-	VA assumed from inclusion criteria	-	+	-	-	-	-	-	-	
Matsuba & Jan (2006)	MDT: psychologist, trained therapist	-	-	-	VA, LV	MRI subgroup	+	-	+	+	+	+	+	
Fazzi *et al.* (2007)	NR	+	+	+	VA, VF, CS, stereo	MRI	+	cog, VP	+	+	+	-	-	
Khetpal & Donahue (2007)	MDT: paediatric ophthalmologist, paediatrician, paediatric neurologist	-	+	+	VF	CT/MRI	-	-	+	+	+	-	+	behaviour problems
Ghasia *et al.* (2008)	NR	+	+	+	VA, VF	-	+	-	-	+	-	-	-	
Ferziger *et al.* (2011)	eye exam by ophthalmologist	+	+	+	VA, VF, LV	-	-	-	+	+	+	-	-	
Good *et al.* (2012)	NR	-	-	-	VA assumed from inclusion criteria	-	+	-	-	-	-	-	-	
Weinstein *et al.* (2012)	NR	+	-	+	VA, VF, stereo	MRI	+	attempted	+	+	+	+	-	ASD, ADHD
Bosch *et al.* (2014a)	paediatric ophthalmologist with assistance from orthoptist	+	+	+	VA, VF	MRI subgroup	-	attempted	+	-	+	-	+	
Bosch *et al.* (2014b)	paediatric ophthalmologist	+	+	+	VA, VF	-	-	attempted	+	-	+	-	-	hypoglycaemia

	Eye examination*								Co-occurring conditions in the sample					
Article ID	**Assessed by**	**fundus exam**	**refraction**	**ocular motility/alignment**	**visual deficits assessment**	**neuroimaging**	**visual electrophysiology included (at least subgroup)**	**cognitive/ neuropsychological assessment**	**intellectual disability/ developmental**	**movement disorder**	**seizure disorder**	**hydrocephalus/ myelomeningocele**	**Hearing impairment**	**Other conditions**
Cavascan *et al.* (2014)	NR	-	-	-	VA	not reported in detail	+	-	+	-	+	+	-	
Chong & Dai (2014)	MDT: paediatric ophthalmologist, paediatrician, optometrist, physiotherapist, speech and language therapist	+	+	-	VA, VF	-	-	-	+	-	-	-	-	
Geldof *et al.* (2015)	"trained researchers", orthoptist	-	+	+	VA, VF, CS, VP, history taking	-	-	cog, VP	+	-	-	-	-	behaviour problems
Mezer *et al.* (2015)	ophthalmologist	-	+	+	VA assumed from inclusion criteria	MRI	-	-	-	-	-	-	-	
Binder *et al.* (2016)	NR	+	-	+	LV	-	-	-	-	-	+	+	-	
Kemmanu *et al.* (2016)	eye exam by ophthalmologist, trained field workers	+	+	-	VA	-	-	-	-	-	-	-	-	
Özturk *et al.* (2016)	ophthalmologist, orthoptist	+	+	+	VA	MRI	-	-	+	-	+	+	-	

*****Assessments only coded as conducted if explicitly listed in the method section of the study, not coded if they were merely listed in the inclusion criteria of the sample

MDT: Multidisciplinary team
NR: Not reported

VA: visual acuity
VF: visual fields
LV: low vision assessment
CS: contrast sensitivity
Stereo: stereopsis

+: yes
-: no

cog : cognitive assessment
VP: neuropsychological visual perceptual assessment

ASD: Autism spectrum disorder
ADHD: attention deficit hyperactivity disorder
DCD: developmental coordination disorder
PDD: pervasive developmental disorder