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Visual and cognitive profiles in children with and without cerebral visual impairment

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Abstract
Reliable differentiation of visual-perceptual difficulties in children with and without cerebral visual impairment (CVI) can often pose a diagnostic challenge. We, therefore, assessed the visual-perceptual profile in 94 children with and 77 children without suspected CVI between the ages of 8 and 17 years in a non-clinical setting, using a screening questionnaire and standardized visual-perceptual tests. Children with suspected CVI reported more frequently greater visual difficulties, had lower visual acuity, and were significantly impaired in visual search tests, in visual form and object perception, in visual space perception, and in visual text processing. There were no significant differences between groups in stereopsis, fixation stability, motility, horizontal saccadic eye movements, and convergence and accommodation. Cognitive performance in auditory attention and verbal short-term and working memory was similar in both groups. Our results indicate that the use of an appropriate questionnaire and specific visual-perceptual tests enables valid diagnostic detection of CVI. The additional use of cognitive tests also allows differentiation between primary and secondary impairments in visual perception.

Keywords
Assessment, cerebral visual impairment, cognition, reading, visual perception

Introduction

The term ‘cerebral visual impairment’ (CVI) is widely used as an umbrella term for a great variety of visual difficulties due to dysfunction of the central visual system (for comprehensive reviews, see Philip & Dutton, 2014; Zihl & Dutton, 2015). Since this diagnostic term is mostly used for children and adolescents with cerebral visual disturbances, Maino (2012) suggested paediatric CVI (PCVI) as a term to differentiate it from CVI in adults.

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A diagnostic classification has not yet been established and there are no valid diagnostic criteria for CVI (Ravenscroft, 2016). For this reason, also secondary visual difficulties, for example, due to cognitive impairments, are often attributed to CVI, even if visual perception is not specifically affected. Children with less severe CVI may not show abnormal visual acuity and (significant) visual field defects may be absent (Fazzi et al., 2007; van Genderen et al., 2012). In addition, the cause of the cerebral impairment of visual perception often cannot be reliably detected by brain imaging methods. For the majority of adults with a CVI, the cause is local brain damage (e.g., stroke) that can be reliably demonstrated on imaging (Zhang et al., 2006). In contrast, the most common cause of CVI is rather diffuse brain injury (hypoxic-ischaemic encephalopathy, periventricular leukomalacia [PVL], hypoxia; Zihl & Dutton, 2015, p. 106) that cannot always be (or is not always) reliably depicted in brain imaging (e.g., Merabet et al., 2017; Ospina, 2009). Furthermore, CVI may also result from genetic causes (Bosch et al., 2016). But even if a pathological change in the brain, that is, the *C* in CVI, can be detected by imaging methods, the investigation of the *V I*, that is, the consequences for visual perception, must be carried out independently in any case. Because of these various difficulties, Boot et al. (2010) have recommended a functional rather than an anatomical approach to address CVI.

On the basis of an extensive and detailed analysis of the criteria for CVI used in the scientific literature, Sakki et al. (2018) have proposed the following definition as a consensus. 'CVI is a verifiable visual dysfunction which cannot be attributed to disorders of the anterior visual pathways or any potentially co-occurring ocular impairment' (p. 431). From a neuropsychological perspective, visual-perceptual difficulty should, in addition, not be fully or sufficiently validly explained by other factors such as impaired cognitive function. This seems important because ~ 50% of children with CVI have difficulties in cognitive domains, especially attention, memory and executive function (Bathelt et al., 2018; Das et al., 2007; Martin et al., 2016; Tadic et al., 2009), which are essential for visual learning processes and for the formation of visual experiences and visual knowledge (Zihl & Dutton, 2015, p. 36). Therefore, although the approach of our study also follows the diagnostic strategy of 'exclusion', a more comprehensive diagnostic assessment may support a more valid characterization of the main features of CVI. The combined inclusion of visual and cognitive functions in the assessment hopefully allows a valid individual diagnostic characterization of a child with (suspected) CVI (Lueck et al., 2019).

Based on the suggestion put forward by Boot et al. (2010), we based our diagnostic approach on visual perception problems that are often reported by children with suspected CVI or by their parents in everyday life situations and in school. In a pilot study on 42 children with (suspected) CVI, we found four main categories of visual impairment: (1) difficulties with global visual processing of scenes (overview, visual exploration and visual search), (2) deficits in form and object perception, (3) deficits in visuospatial functions, and (4) difficulties with reading (text processing).

Difficulties in visual exploration/visual search have already been reported by previous authors (Fazzi et al., 2007; Netelenbos & Van Rooij, 2004; Salati et al., 2002). Children with these abnormalities typically need more time to search through a stimulus template and are more likely to neglect stimuli. They show difficulties with parallel visual processing of a scene and the spatial (and possibly also temporal) integration of visual information. As a result, they find it difficult to grasp scenes as a whole and to get a complete and quick overview. Their visual search pattern is characterized by a serial procedure, with many fixations and, as a result, a significantly increased time requirement compared to healthy children (Zihl & Dutton, 2015, p. 92). Similar difficulties may arise with visual word processing at the pre-semantic level. Children with this subtype of developmental visual dyslexia typically are able to discriminate letters and can identify/recognize them, but are unable to combine the individual letters into words, that is, to

process words holistically (Goldstein-Marcusohn et al., 2020). Impaired parallel search and holistic word processing can also occur in combination (Jones et al., 2008; Sireteanu et al., 2008; Zihl & Dutton, 2015, p. 271). Difficulties in visual form and object perception have also been reported in subjects with CVI (Fazzi et al., 2007; Van der Zee et al., 2019). Finally, we also examined visual space perception, as this can also be affected in children with CVI (Fazzi et al., 2007; Zihl & Dutton, 2015, p. 82). In the domain of cognition, we assessed auditory attention and verbal short-term and working memory. Because of possible interference effects with visual difficulties, we did not assess visual attention and visual short-term and working memory. In addition, we used a modified standardized question inventory for screening and assessing visual difficulties in everyday life activities (Gorrie et al., 2019; Zihl & Dutton, 2015, pp. 147–155).

The main aim of our study is to answer the question of whether a clinically useful distinction can be made between children with and without CVI based on a specific questionnaire and selected visual test procedures. This methodological approach seems sensible for two reasons. On the one hand, not every central nervous dysfunction is associated with visual disturbances; on the other hand, in many cases, there is insufficient evidence for the involvement of brain structures; however, subjects nevertheless may report visual difficulties.

Methods

Study design

In this single-centre study, children have been registered in our counselling centre initiated by parents by letter, phone, or e-mail, on advice from special education teachers and school psychologists (50%), paediatricians (15%), occupational and speech therapists (15%), ophthalmologists and orthoptists (10%), or early intervention specialists (10%). Based on the results of the standardized CVI interview (see below) and available medical evidence, subjects who met the criteria for a possible CVI were invited to the examination.

Ethical considerations

The research study was carried out according to the World Medical Declaration of Helsinki and approved by the Ethics committee of the Faculty of Psychology and Educational Sciences, University of Munich (LMU) (approval number: 06_a_2015). After a detailed explanation of the purpose of the research study, subjects and their parents gave written consent for the voluntary participation and the use of anonymized data for scientific purposes. It was explicitly stated that withdrawing this consent would not result in any disadvantages. The number of test procedures and thus the duration of the examination were chosen so that subjects would not experience excessive stress and compliance was sufficiently given.

Setting

The examination took place in a quiet room under normal daylight conditions in the Centre for the Visually Impaired in Unterschleißheim (Bavaria, Germany). Examinations were carried out between 9 a.m. to 1 p.m. An individual examination took on average 110 min (range: 90–160 min), including breaks. The subjects could decide for themselves when they wanted to have a break. There were breaks of 5–10 min after every 30 min of testing, and more often if necessary. Prior to the study, the subjects were informed that they could refuse individual tasks if they found them too difficult or uncomfortable.

Table 1. CVI Questionnaire.

Items
Global perception and visual search
(1) Does your child have difficulty to avoid people or objects?
(2) Does your child bump against obstacles?
(3) Does your child have difficulty to find a known person among other people?
(4) Does your child have difficulty to find a special toy among many toys?
(5) Does your child have difficulties to find a specific piece of clothing?
Visuospatial orientation
(1) Does your child have difficulties to find a common path?
(2) Does your child have difficulty to orient itself in a familiar place?
(3) Does your child have difficulty to orient itself in a familiar supermarket?
Visually guided activities
(1) Does your child have trouble catching a ball?
(2) Does your child have trouble walking on uneven ground?
(3) Does your child miss or knock over objects when reaching for them?
(4) Does your child have difficulties using tools, e.g., when playing?
Reading
(1) Does your child omit letters at the beginning or end of longer words?
(2) Does your child have trouble grasping longer words as a whole?

Scoring: never (0), sometimes (1), frequently (2).
CVI: cerebral visual impairment.

Study population

Between January 2017 and February 2022, we received 456 inquiries for examination of subjects suspected of children suspected to have CVI. Based on the available medical evidence and the results of the interview using the CVI Questionnaire, an appointment for an examination was finally arranged with 171 subjects were examined. The decision in favour of a neuropsychological examination for the detection of CVI was based on the following inclusion criteria: (1) outcome of the CVI Questionnaire (see Table 1); inclusion criterion was a score of $\geqslant 2$ in at least one of the four categories, (2) ophthalmological/orthoptic indications of a visual impairment unexplained by impaired peripheral visual system, and (3) medical diagnoses, if available, for example, premature birth, oxygen problems during or after birth, imaging evidence (PVL, etc.). Exclusion criteria were: (1) a total score of <2 in the CVI Questionnaire, and (2) sufficient explanation of the visual difficulties through peripheral causes of the visual system by ophthalmological examination and/or through cognitive or behavioural problems (e.g., global cognitive developmental delay, attention-deficit/hyperactivity disorder [ADHD]). Subjects who fulfilled the inclusion and exclusion criteria were assigned to the CVI group (CVI+), while subjects who did not fulfil the inclusion criteria, but fulfilled the exclusion criteria, were assigned to the control group (CVI−). In eight subjects with a medical diagnosis, questionnaire scores were smaller than required for inclusion in the CVI+ group (overall score <2); these subjects were, therefore, assigned to the CVI− group.

Data collection

After reviewing the available medical documents and the outcome of the CVI Questionnaire, 94 subjects met the criteria for CVI+; the control group (CVI−) consisted of 77 subjects. For both groups, we collected demographic variables (age, gender) and information on additional diagnoses. Visual difficulties were recorded with the CVI Questionnaire, which consisted of 14 items (Table 1).

Table 2. List of tests and performance variables.

Test	Performance variables
Dot Cancellation	Speed (time, in s) Accuracy (correctly identified items)
modified Teddy Bear Cancellation Test	Speed (time, in s) Accuracy (correctly identified items)
Selective Visual Attention (BVN/NPS 5-11)	Accuracy (correctly identified items, 1 min)
Form Constancy (FEW-2/JE)	Correctly matched items
Figure-Ground (FEW-2/JE)	Correctly matched items
Visual Closure (FEW-2/JE)	Correctly matched items
Object recognition, naturalistic	Correctly identified items
Object recognition, prototypic	Correctly identified items
Position in space (FEW-2)	Correct responses
Line orientation test (Benton)	Correct responses
Position perception	Correctly matched positions
Reading, words	Correctly read items/time (s)
Reading, text (LDL)	Correctly read words/min
Reading, digits	Correctly read items/time (s)
Digit span forwards (WISC-IV)	Correct responses
Digit span backwards (WISC-IV)	Correct responses
Selective Auditory Attention (BVN/NPS 5-11)	Correctly identified items
Strengths and Difficulties Questionnaire (SDQ-E)	Emotional problems, conduct problems, peer problems, hyperactivity, total difficulties, prosocial behaviour

Assessment procedures

Assessment included visual and cognitive tests and a mental health questionnaire (see Table 2). Assessment of visual perception included visual search, visual form and figure-ground discrimination, Gestalt perception, visual space perception, visual object recognition, and visual text processing/reading. Prior to the assessment, subjects were familiarized with the respective test procedure in a short exercise. Tests were administered in a counterbalanced order to avoid systematic presentation order effects; in addition, subjects were allowed as many breaks as they wished.

Visual search. For the assessment of visual search performance, we used three different tasks: a dot cancellation task (see Zihl & Dutton, 2015, p. 171; fig. 6.4c), a modified version of the Teddy Bear Cancellation Test (Laurent-Vannier et al., 2006; Unterberger, 2016), and the BVN 5-11 test (Bisiacchi et al., 2005; German version: Kaufmann et al., 2008). In the dot cancellation task, subjects were asked to cross out 20 black dots (diameter: 6 mm) distributed randomly on a white sheet of paper (size: 200 × 300 mm; distance between dots: 40–70 mm). This test form was chosen in order to prevent unfavourable influences by differentiating between forms. In the modified version of the Teddy Bear Cancellation Test, targets are 15 Teddy bears (line drawings, size: 20 × 15 mm) which were randomly distributed among 60 distractors of the same size (cars, birds, shoes, umbrellas, dolls, gloves, and sweets; line drawings) on a white sheet of paper (size: 200 × 300 mm). The distance between items varied between 5 and 15 mm. Subjects are asked to search for the Teddy Bears and to cross them out with a pencil as accurate and fast as possible. For both test procedures, subjects carried out a brief practice task beforehand to familiarize themselves with the respective task. In the BVN 5-11 test, subjects were asked to search for a target form among non-target forms.

This test consists of 10 lines with 8 squares each (size: 17 × 17 mm; distance between squares: 7 mm) with 2 lines (length: 5 mm) of different orientations at different positions. The target figure is in the middle at the top of the template. The distractors differ from the target stimulus in position and orientation of the two lines in the square. Subjects were asked to search as accurately and fast as possible for the target figure and to cross it out; the processing time is 1 min.

Form, Gestalt, and Figure-ground perception were assessed using the figure-matching, the figure-ground discrimination, and the gestalt identification subtests of the developmental test for visual perception (DTVP; Hamill et al., 1993) for children ⩽8 years (FEW-2; Büttner et al., 2008) and ⩾9 years (FEW-JE; Petermann et al., 2012).

Visual space perception. For the assessment of position perception, we used the subtest 'position in space' (FEW-2; Büttner et al., 2008; ⩽8 years) and Benton's Judgement of Line Orientation Test (Benton et al., 1978; ⩾ 9 years).

Object perception. Object perception was assessed using two sets of photographs (Unterberger, 2016). The first set consisted of 12 photographs of real objects ('real objects') taken from Moreno-Martínez and Montoro (2012), the second set consisted of 12 black and white drawings ('prototypical objects') taken from Snodgrass and Vanderwart (1980). For the gradation of complexity, we used the criteria proposed by Snodgrass and Vanderwart (1980); for each complexity (six levels) we selected two items for both sets. Each item was shown on a single page (200 × 300 mm); the drawings were reworked to improve contrast and thickness of lines (6 pt). Each set was shown independently, the order of presentation of single items was at random. Performance was defined as number of correctly identified items by naming the objects, whereby subjects could also paraphrase. The total score was 12 for each set. Normative data were taken from Unterberger (2016).

Visual text processing/reading. Visual word processing was assessed only in children >7 years using a standardized German reading test for sentence reading (LDL; Walter, 2009). Subjects were asked to read a text (2–11 letter words) aloud as correctly and as fast as possible. Reading performance is defined as number of words correctly read in 1 min.

Cognition. Verbal short-term and working memory was assessed using the digit span tests (subtests 'digit spans forward and backward'; WISC, Petermann et al., 2007). In addition we also assessed verbal problem solving (subtest similarities"; WISC IV; Petermann et al., 2007). For the assessment of selective auditory attention, we used a subtest of the BVN/NPS 5-11, Batteria di valutazione neuropsicologica per l'età evolutiva (BVN 5-11 (Bisiacchi et al., 2005; German version: Kaufmann et al., 2008). A text was read to the subjects in which a certain word appears repeatedly. The subjects were asked to knock on the table every time they hear this word.

Other domains. Aspects of mental health were assessed with the Strengths and Difficulties Questionnaire (SDQ; Goodman, 2001; German version: SDQ-Deu; Woerner et al., 2002). The SDQ consists of 25 items, which are related to emotional, social and behavioural difficulties of children and this questionnaire was completed by a parent.

Statistical analysis

Statistical evaluation was performed with IBM SPSS 27. Based on the available normative data, the results of the tests were divided into three groups (average, below average, and above average), corrected by age, to describe the data as well as to compare the distribution of the performance

between the two groups. Due to age differences between subjects, different tests were used for the corresponding age groups; for the statistical evaluation the test results were combined. For statistical comparison of the CVI+ and CVI− subjects, we used the Mann–Whitney *U* test, and for frequencies, the Pearson Chi-square test; the level of significance was calculated two-tailed. In addition, we also calculated partial correlations between the performance in the various visual-perceptual tests and visual far and near acuity corrected for age using Kendall's Tau. Because of the multiple group comparisons, we set the significance level p to $<.01$ to correct the p for consequent effects. Since the subjects could refuse the visual and cognitive tests or could stop if it was too difficult for them, unfortunately the number of participants did not reach 100% in any test. The number of subjects in the different tests differed between 35 (37.2%; position perception) and 86 (91.5%; modified Teddy Bear Cancellation Test) in the CVI+ group, and between 21 (27.3%; copying) and 70 (90.1%; Teddy Bear test) in the CVI− group. Reading performance was only assessed in subjects >7 years; 37 subjects (49.3%) of the CVI+ group ($n=75$) and 45 subjects (71.4%) of the CVI− group ($n=63$) completed the reading test.

The outcome of the CVI Questionnaire in the two groups was not further statistically analysed because the scores only served as inclusion criteria for this study.

Results

Subjects characteristics

Table 3 shows the demographical and clinical data for both groups. Age was similar in both groups; the majority of children were older than 7 years in both groups (CVI+: 79.8%; CVI−: 82.3%) and the two groups did not differ significantly in age ($t=0.44$; $p=.664$). Both groups included more boys than girls (CVI+: 60.6%; CVI−: 68.8%). CVI+ diagnosis was based on brain imaging (PVL) or empirical evidence of a brain developmental disorder in 56% of children; for the remainder of the sample, the diagnosis was based only on the outcome of the CVI Questionnaire. In about one-third of the CVI+ group, the medical diagnosis was hypoxia; in about 12% it was a genetic disorder (microcephaly: five cases, Prader-Willy syndrome: one, Williams syndrome: one). Other brain diseases were central nervous system (CNS) infection in two and occipital stroke in three cases. Cerebral palsy was present in nine subjects.

In the CVI− group, genetic developmental disorders were Asperger syndrome in four, Sturge-Weber syndrome in two, and West syndrome in one subject. Eight children had retinal disorders (ROP: six, retinoblastoma, operated: two). Global developmental delay was more than twice as common in the CVI+ group (~47%) than in the CVI− group (~22%).

CVI Questionnaire

Table 4 shows the outcome of the questionnaire for both groups. Overall, the CVI+ group reported more frequently visual difficulties and achieved almost twice as high scores in all four categories compared to the CVI− group. Interestingly, however, nearly 70% of children in the CVI− group also showed visual difficulties in the categories overview/visual search and reading, ~50% in visuospatial orientation and visually guided activities, and ~40% in reading. Scores in these categories were, however, markedly lower compared to the CVI+ group. Comparing the frequencies of reported difficulties at the highest score of the CVI− group in each category to the frequencies in the CVI+ group adds further evidence that considerably more subjects in the CVI+ group reported such difficulties. Interestingly, in the CVI+ group the rates of spatial orientation difficulties above these values were lowest (~6%), followed by difficulties with visually guided activities (~19%),

Table 3. Demographic and clinical data for CVI+ and CVI− groups.

CVI+ group ($n=94$)				CVI− group ($n=77$)		
Demographic data						
Age		*M* (*SD*)	9.14 (3.04)	Age		9.32 (2.56)
		Range	5–17			5–16
Gender	Female	*n* (%)	37 (39.4%)	Gender	Female	24 (31.2%)
	Male		57 (60.6%)		Male	53 (68.8%)
CVI diagnosis						
Brain		*n* (%)	30 (31.9%)			
Brain developmental disorder			23 (24.5%)			
Suspected				41 (43.6%)		
Diagnoses						
Preterm, with hypoxia		*n* (%)	21 (22.3%)			
Term, hypoxia			7 (7.5%)			
Genetic disorders			11 (11.7%)			7 (9.19%)
Epilepsy			4 (4.3%)			7 (9.1%)
Brain tumour (operated)			7 (7.5%)			
Other brain diseases (e.g. infectious or ischemic diseases)			5 (5.3%)			
Global cognitive developmental delay			42 (46.8%)			17 (22.1%)
Developmental motor disorder (fine motor skills)			11 (11.7%)			11 (14.28)
Cerebral paresis			9 (9.6%)			
Reading/writing difficulties			5 (5.3%)			
ADHD						29 (37.7%)
Retinal disorders			00			8 (10.4%)
No additional diagnosis			15 (16.0%)			
No clinical information available			39 (41.5%)			

SD, ranges, and percentages in parentheses. Diagnoses: multiple entries possible.
CVI: cerebral visual impairment; *SD*: standard deviation; ADHD: attention-deficit/hyperactivity disorder.

difficulties with text processing (~22%), and difficulties with visual search/visual overview (~39%). In contrast, the corresponding frequency values were considerably lower in the CVI− group in all categories (<9%), apart from the frequency of spatial orientation difficulties (5.2%) (see Table 4).

Ophthalmological and orthoptic data

Table 5 summarizes the ophthalmological and orthoptic examination results for both groups.

Visual acuity. Mean corrected far and near binocular acuity values were slightly lower in the CVI+ group. Visual far and near acuity was >0.80 in ~69% in the CVI+ group; the corresponding percentages for the CVI− group were ~82% and ~ 74%. Both far and near visual acuities differed significantly between the two groups (far visual acuity: $U=2769.00$; $p=.009$; near visual acuity: $U=2648.500$; $p=0.001$).

Stereopsis (age-corrected) was not significantly more often impaired in CVI+ subjects (~51%) than in CVI− subjects (~34%); $\chi^2(1)=4.84$; $p=.028$.

Table 4. Results of the CVI Questionnaire for the CVI+ group ($n=94$) and the CVI− group ($n=77$).

(a)

	CVI+ *n* (%)	CVI− *n* (%)
Overview/visual search	91 (94.8%)	52 (67.5%)
Visuospatial orientation	81 (86.2%)	38 (49.5%)
Visually guided activities	83 (88.3%)	39 (50.6%)
Reading	34 (91.9%)	31 (40.3%)

(b)

	CVI+ *M* (*SD*), range	CVI− *M* (*SD*), range
Total scores	13.6 (3.5), 5–21	7.6 (2.3), 0–13
Overview/visual search	4.3 (4.2), 5–10	2.2 (1.0), 0–5
Visuospatial orientation	2.7 (1.2), 0–6	1.6 (1.0), 0–4
Visually guided activities	3.2 (1.6), 0–7	1.8 (1.2), 0–5
Reading	3.2 (1.6), 0–6	1.9 (1.0), 0–3

(c)

	CVI− Max. score/f (%)	CVI+ >f/max. score (%)
Overview/visual search (10)	4/6 (7.8%)	37 (39.4%)
Visuospatial orientation (6)	4/4 (5.2%)	06 (6.4%)
Visually guided activities (8)	4/7 (9.0%)	18 (19.1%)
Reading (4)	4/7 (9.1%)	21 (22.3%)

For reading, corresponding numbers were 37 in the CVI+ and 45 in the CVI− group. (a) Frequencies and percentages (in parentehses) refer to scores ⩾2; (b) Means, standard deviations, and ranges for scores for CVI+ and CVI− groups. (c) Frequencies (% in parentheses) of reported difficulties above the highest scores (max. scores) in the respective category in the CVI− and the CVI+ groups. Note the reversed order for CVI+ and CVI− in (c).
CVI: cerebral visual impairment; *SD*: standard deviation.

Visual field. Nearly all subjects (~93%) in the CVI+ group had a normal visual field. Five subjects suffered from a bilateral homonymous constriction in the peripheral visual field, but visual field sparing was ⩾30° in all cases. Two subjects showed incomplete homonymous right-sided hemianopia (visual field sparing: 10° and 14°). In the CVI− group, two subjects (~3%) with retinal blastoma (operated) showed a slight concentric visual field constriction in both eyes beyond 20° eccentricity.

Oculomotor functions. In the cover test, more subjects in the CVI+ group presented with ocular deviation. In this group, esophoria or esotropia was present in 27 subjects (28.7%), and exophoria or exotropia in 15 subjects (16%). In contrast, in the CVI− group, 10 subjects (~13%) presented with esophoria or esotropia, and 9 subjects (~12%) with exophoria or exotropia. The difference in the frequencies of the two groups was statistically significant, $\chi^2(1)=14.69$; $p=.005$. Ocular motility ranges were mostly preserved in both groups, and frequencies did not differ significantly, $\chi^2(1)=2.04$; $p=.153$. Horizontal saccadic eye movements were normal in the majority of subjects

Table 5. Ophthalmological and orthoptic data.

Variable		CVI+ ($n=94$)	CVI− ($n=77$)
Far acuity	*M* (*SD*)	0.80 (0.25)	0.90 (0.20)
	Range	0.1–1.0	0.3–1.3
⩾1.00	*n* (%)	44 (46.8%)	46 (59.7%)
0.80–0.90		21 (22.3%)	17 (22.1%)
0.50–0.70		17 (18.1%)	11 (14.3%)
<0.50		12 (12.8%)	3 (3.9%)
Near acuity	*M* (*SD*)	0.80 (0.27)	0.90 (0.20)
	Range	0.1–10	0.3–1.3
⩾1.00	*n* (%)	51 (54.3%)	41 (53.2%)
0.80–0.90		14 (14.9%)	16 (20.8%)
0.50–0.70		15 (15.9%)	14 (18.2%)
<0.50		12 (12.8%)	06 (07.8%)
Stereopis (age-corrected)			
Normal	*n* (%)	46 (48.9%)	51 (66.2%)
Impaired		48 (51.1%)	26 (33.8%)
Visual field			
Normal	*n* (%)	87 (92.6 %)	75 (97.4%)
Impaired		7 (07.4%)	2 (02.6%)
Strabismus			
Esophoria	*n* (%)	7 (7.4%)	7 (9.1%)
Esotropia		20 (21.3%)	3 (3.9%)
Exophoria		4 (4.3%)	5 (6.5%)
Exotropia		11 (11.7%)	4 (5.2%)
Ocular motility			
Normal	*n* (%)	86 (91.5%)	65 (84.4%)
Impaired		8 (8.5%)	12 (15.6%)
Saccadic eye movement (horizontal)			
Normal	*n* (%)	85 (90.4%)	76 (98.7%)
Impaired		9 (9.6%)	1 (1.3%)
Pursuit eye movements (horizontal)			
Normal	*n* (%)	37 (39.4%)	57 (74.0%)
Impaired		57 (60.6%)	20 (26.0%)
Fixation			
Normal	*n* (%)	67 (71.3%)	68 (88.3%)
Instable		21 (22.3%)	09 (11.7%)
Nystagmus		6 (6.4%)	
Convergence			
Normal	*n* (%)	77 (80.2%)	68 (88.3%)
Impaired		19 (19.8%)	9 (11.7%)
Accommodation (age-corrected)			
Normal	*n* (%)	71 (75.5%)	68 (88.3%)
Impaired		23 (24.5%)	9 (11.7%)

CVI: cerebral visual impairment; *SD*: standard deviation.

in both groups (>90%), but CVI+ subjects showed significantly more often dysmetria (~10% vs ~1%); $\chi^2(1)=5.34=5.34$; $p=.021$. One subject had signs of ocular apraxia. Horizontal pursuit eye movements were impaired in about 60% of CVI+ subjects, but only in ~26% in subjects without

CVI; the difference was statistically significant, $\chi^2(1)=20.97$; $p<.001$. About 70% of subjects with CVI had normal fixation; in the remaining sample fixation was either unstable or fixation nystagmus was present. In the CVI− group, ~88% of subjects had normal fixation; whereas ~ 12% showed an instable fixation, but no subject presented with fixation nystagmus. The difference in fixation impairments between the groups was not statistically significant, $\chi^2(2)=7.02$; $p=.030$. Convergence and age-corrected accommodation were more often impaired in the CVI+ group (~20% and 25%, respectively) than in the CVI− group (~12%), but differences were not statistically significant, larger χ^2 $(1)=4.690$; $p=.030$.

Visual perception

The outcome of the assessment of visual-perceptual abilities for the CVI+ and the CVI− groups is shown in Table 6.

Visual search. The main characteristic of visual search performance in the CVI+ group was a significant slowing in all three tests, with the highest percentage of subjects in the dot cancellation test (~81%), followed by the modified Teddy Bear Cancellation Test (~74%). Accuracy was much better in the two search tests (dot cancellation test: ~90%, Teddy Bear test: ~65% >cut-off values). In the CVI− group, about half of the subjects showed reduced speed in the dot cancellation task, and about 25% in the Teddy Bear test. Concerning accuracy, about 97% of subjects performed above the cut-off values. Overall, CVI+ subjects performed worse in the visual search tests than CVI− children, in terms of both speed and accuracy. CVI+ subjects performed worse in all search tests, both in accuracy and in speed ($U=1195.0$–2018.0, $p<.001$), except for accuracy in the dot cancellation test ($U=2563.0$, $p=.045$). According to the number of correctly identified targets per second which was used as performance parameter (n targets−n errors/s), 81.4% of subjects in the CVI+ group and 24.6% of subjects in the CVI− group performed below the cut-off value; the difference was statistically significant ($U=1195.0$; $p<.001$).

Figure-ground, form, and gestalt perception. Nearly half of the subjects in the CVI+ group had difficulties with these tasks; about 44% performed below the cut-off value in form perception, ~64% in gestalt and ~51% in figure-ground perception. In contrast, subjects in the CVI− group performed significantly better in these tasks, where only ~14% performed below the cut-off value in form and figure-ground perception, and ~19% in gestalt perception (lowest U-value $=1791.00$; $p<.001$).

Object perception. In the naturalistic object test, ~25% of subjects in the CVI+ group performed below the corresponding cut-off value, and ~34% in the prototypical object test. In contrast, only ~11% of the CVI− group performed below the cut-off value in the naturalistic version, and ~5% in the prototypic version of the test. Thus, subjects of the CVI+ group found both object visual recognition tests more difficult. While performance for prototypical objects differed significantly between groups ($U=1892.00$; $p=.009$), differences in natural object recognition did not reach statistical significance ($U=2271.00$; $p=.038$).

Visual space perception. In both, position and line matching tests, more subjects in the CVI+ group performed below the corresponding cut-off values (~58% and ~ 71%, respectively) than subjects in the CVI− group (~13% and ~17%, respectively). The differences were statistically significant ($U=227.50$–1525.50; $p<.01$).

Copying. As in the tests on visual form and gestalt perception and on visuospatial tasks, more subjects in the CVI+ group (~77%) performed below the cut-off scores than subjects in the

Table 6. Frequencies (and respective percentages, in parentheses) and statistical results of values <cut-off for visual-perceptual, cognitive, and mental health data for the CVI+ and CVI−groups.

Variable	CVI+	CVI−	Mann-Whitney *U*; significance	Effect size (*r*)
Visual perception				
Visual search				
Dot cancellation (speed)	68 (80.6%, *n* = 84)	33 (49.3%; *n* = 67)	*U* = 1892.00; *p* < .001	.34
Dot cancellation (accuracy)	10 (11.9%; *n* = 84)	02 (02.9%; *n* = 67)	*U* = 2563.00; *p* = .045	.16
Dot cancellation (speed and accuracy)	69 (82.1%; *n* = 84)	35 (52.2%; *n* = 67)	*U* = 1944.50; *p* < .001	.33
Modified Teddy Bear Cancellation Test (speed)	64 (74.4%; *n* = 86)	17 (24.6%; *n* = 69)	*U* = 1369.00; *p* < .001	.52
Modified Teddy Bear Cancellation Test (accuracy)	30 (34.9%; *n* = 86)	2 (2.9%; *n* = 69)	*U* = 2018.00; *p* < .001	.39
Modified Teddy Bear Cancellation Test (speed and accuracy)	70 (81.40%; *n* = 86)	17 (24.6%; *n* = 69)	*U* = 1195.00; *p* < .001	.58
Selective visual attention	42 (58.3%; *n* = 72)	17 (24.3%; *n* = 70)	*U* = 1392.00; *p* < .001	.31
Form constancy	37 (44.6%; *n* = 83)	10 (14.5%; *n* = 69)	*U* = 1791.00; *p* < .001	.35
Visual closure	52 (64.2%; *n* = 81)	14 (19.2%; *n* = 73)	*U* = 1524.00; *p* < .001	.46
Figure-ground	41 (50.6%; *n* = 81)	11 (13.9%; *n* = 74)	*U* = 1558.50; *p* < .001	.45
Object perception (naturalistic)	20 (24.4%; *n* = 82)	07 (10.8%; *n* = 65)	*U* = 2271.00; *p* = .038	.17
Object perception (prototypic)	28 (34.1%; *n* = 82)	04 (05.1%; *n* = 65)	*U* = 1892.00 *p* < .001	.33
Position perception	46 (58.2%; *n* = 79)	08 (13.3%; *n* = 60)	*U* = 1525.50; *p* < .001	.38
Space perception	25 (71.4%; *n* = 35)	06 (17.1%; *n* = 35)	*U* = 227.50; *p* < .001	.59
Reading, words	16 (43.2%; *n* = 37)	13 (28.9%; *n* = 45)	*U* = 592.00; *p* = .003	.33
Cognition				
Selective auditory attention	20 (39.3%; *n* = 51)	09 (23.1%; *n* = 39)	*U* = 834.00; *p* = .044	.21
Verbal short-term memory	26 (56.5%; *n* = 46)	12 (44.4%; *n* = 27)	*U* = 568.50; *p* = .496	.08
Verbal working memory	14 (34.1%; *n* = 41)	05 (20.0%; *n* = 25)	*U* = 416.00; *p* = .050	.24
Verbal problem-solving skills			*U* = 19.50; *p* = .905	.02
Mental health				
Total scores			*U* = 642.50; *p* = .011	.28
Emotional problems	12 (27.3%)	21 (51.2%)	*U* = 651.50; *p* = .011	.28
Conduct problems	06 (13.6%)	18 (43.9%)	*U* = 632.00; *p* = .003	.33
Peer problems			*U* = 810.00; *p* = .375	.10
Hyperactivity	08 (18.2%)	06 (14.6%)	*U* = 681.50; *p* = .030	.24
Prosocial behaviour			*U* = 857.00; *p* = .539	.07

CVI: cerebral visual impairment.
For details, see text

CVI− group (~51%), but interestingly only half of the subjects in this group were not impaired in this task. The difference though was not statistically significant ($U = 352.50$; $p = .115$).

Visual word processing (reading). In the visual word processing (reading) test, more subjects in the CVI+ group (~43%) performed below the cut-off score than in the CVI− group (~30%). The difference between the two groups was statistically significant ($U = 592.00$; $p = .003$).

Table 7. Correlations between visual far and near acuity and frequencies of CVI+ subjects in visual-perceptual tests.

Variable	*n*	Far acuity (τ)	Near acuity (τ)
Dot cancellation, speed	72	−0.001, $p = .496$	0.38, $p = .038$
Teddy Bear, speed	72	0.13, $p = .135$	0.07, $p = .295$
BVN/NPS 5-11 Selective visual attention	72	0.08, $p = .258$	−0.03, $p = .412$
Figure-ground	77	0.38, $p <$ **.001**	0.37, $p <$ **.001**
Visual Closure	77	0.14, $p = .109$	0.16, $p = .161$
Form Constancy	77	0.25, $p = .014$	0.24, $p = .020$
Object perception, naturalistic	81	0.27, $p =$ **.008**	0.37, $p <$ **.001**
Object perception, prototypic	81	0.40, $p <$ **.001**	0.38, $p <$ **.001**
Position perception	82	0.14, $p = .112$	0.07, $p = .273$
Space perception	35	0.06, $p = .745$	0.17, $p = .348$
Reading	32	0.09, $p = .310$	0.14, $p = .221$

Significant correlations ($p < .01$) in bold.

Visual acuity and visual-perceptual performance. Table 7 summarizes the correlations between far and near visual acuity and performance in visual-perceptual tests. Significant correlations were only found for figure-ground and for object perception.

Cognition

In the selective auditory attention task, about twice as many subjects in the CVI+ group (~39%) performed below the cut-off score than in the CVI− group (~23%). Short-term verbal memory performance was impaired in more than 40% of subjects in both groups, with more impaired subjects in the CVI+ group (CVI+ group: ~56%; CVI− group: ~44%). For verbal working memory, the number of impaired subjects was lower in both groups (CVI+ group: ~34%, CVI− group: 20%). We found no statistically significant difference for either cognitive task performance ($U = 416.000–835.000$; $p \geq .04$).

Summarizing the outcome of our study, the two groups differ significantly in some categories of the CVI Questionnaire as well as in the visual tests. In the Questionnaire, subjects in the CVI+ group reported much more frequently difficulties in three out of four categories, that is, overview/visual search, visually guided activities and reading. Ranking the outcomes of visual tests based on effect sizes, space perception, performance (speed and accuracy) in the Teddy Bear cancellation test, visual closure and figure-ground perception achieved effect sizes between .59 and .40, while effect sizes in the remaining tests varied between .38 and .33 (see Table 6). Thus, effect sizes ranged from strong (space perception and Teddy Bear cancellation tests; >.50) to medium ($.30 < r < .50$) in the other tests. For object perception and for the cognitive tests, the effect size was weak ($.10 < r < .30$).

Mental health

Interestingly, contrary to the results reported so far, more subjects in the CVI− group than in the CVI+ group showed high SDQ scores, not only for the total score (~56% vs ~32%) but also for the categories emotional (~51% vs ~27%) and behavioural problems (~44% vs ~14%). However,

in the category 'prosocial behaviour', the percentages were similar in both groups (CVI+ group: ~18%; CVI− group: ~15%). The differences in total scores and scores for emotional and behavioural problems reached statistical significance ($U = 632.0–510.0$; $p = .011$); in contrast, scores in prosocial behaviour did not differ significantly ($U = 857.0$; $p = .539$). Effect sizes were weak ($.1 < r < .3$) for all variables, except for conduct problems ($r = .33$; see Table 6).

Discussion

The main outcome of this study is that subjects with a diagnosis of CVI according to our chosen criteria reported more frequently and greater visual difficulties, showed slightly, but significantly lower (far and near) visual acuity values, presented significantly more frequent with ocular deviation, and exhibited significantly more often impaired horizontal smooth pursuit eye movements compared to subjects without such diagnosis. Visual-perceptual performance was significantly impaired in visual search, figure-ground and form and gestalt perception, prototypical object perception, visual space perception, and visual text processing. Figure-ground and object perception, but not reading performance, are significantly correlated with visual acuity. There were no significant differences between groups in stereopsis, fixation stability, motility, horizontal saccadic eye movements, and convergence and accommodation. In addition, performance in the natural object perception test and in figure copying did also not differ significantly. Cognitive performance in auditory attention and verbal short-term and working memory was similar in both groups. Interestingly subjects of the CVI− group showed significantly more emotional and behavioural abnormalities in the SDQ than subjects of the CVI+ group, but both groups scored similarly concerning prosocial behaviour.

Difficulties with global visual perception (overview) and visual search have been frequently reported in CVI (Fazzi et al., 2007; Netelenbos & Van Rooij, 2004; Salati et al., 2002; Zihl & Dutton, 2015, p. 92). These difficulties may be interpreted in terms of impaired parallel visual processing and spatial integration, resulting in a restriction of the field of attention, which is also a characteristic feature of (mild) Balint syndrome (Philip et al., 2016). Impaired global perception can cause severe difficulties with activities in school and in everyday life because it often hinders the full and timely perception of the current physical and social environment. Difficulties with reading in our subjects may also be explained by impaired spatial (and perhaps temporal) integration of visual text processing, resulting in a letter-by-letter strategy of processing. The resulting slowdown in the reading process does not only impair reading comprehension but also often leads to more rapid fatigue due to the increased need for concentration. Impaired spatial localization and line orientation may result in difficulty copying and drawing especially geometric figures, which can have an unfavourable effect, especially at school. The same holds true for difficulties with form and object perception, but the familiar context in everyday life situations may be helpful. We did not assess contrast vision and crowding, because no subject of either group reported typical phenomena of 'visual blurring' and/or 'merging' of adjacent lines or figure details during visual assessment. Interestingly, we only found significant correlations with visual acuity for performance in the figure-ground and object perception tests. This outcome is in agreement with other studies showing that visual acuity may be preserved in subjects with CVI (Chandna et al., 2021; van Genderen et al., 2012). None of our subjects reported difficulties seeing the test templates, but we cannot rule out, that reduced visual acuity may have contributed to these below-average test performances. On the other hand, one might also assume that visual acuity, figure-ground discrimination, and object recognition were affected jointly but independently. However, these results also show that good visual acuity does not necessarily mean that higher levels of visual perception are intact.

Although the CVI+ and CVI− groups did not differ significantly in cognitive performance, we cannot rule out that reduced attention and working memory may have influenced some results to a higher degree in the CVI+ group. Furthermore, fixation, smooth pursuit eye movements, and stereopsis also depend on maturational stage (Heinbuck & Hershberger, 1989; Papageorgiou et al., 2014; Sinno et al., 2020), and abnormalities may thus not be specific to CVI. For this reason, we did not use oculomotor abnormalities as inclusion criteria for CVI+, but have included them as additional variables.

Vision impairment can have an impact on mood in children, as they may show more often symptoms of depression and anxiety (Li et al., 2022). Although we did not address depression and anxiety specifically, subjects in our CVI+ group did not show higher abnormal scores for emotional and behavioural problems.

In conclusion, our data show that a careful medical history taking, a questionnaire appropriate to assess specific visual problems, a thorough ophthalmological and orthoptic examination and, if possible, a neurological examination, and standardized test procedures to assess visual perception and cognitive function should be part of the diagnostic standard. The combination of this multidisciplinary assessment appears a viable approach to detect and characterize CVI, and to separate primary and secondary visual disturbances with satisfactory reliability (see Boonstra et al., 2022; McConnell et al., 2020, for comprehensive reviews). A questionnaire alone, no matter how extensive it may be, can in no way replace the quantitative examination of visual and cognitive functions (see also van Genderen et al., 2012). However, this does not mean that all possible or conceivable visual and cognitive functions have to be assessed in every subject with (suspected) CVI in order to obtain a complete profile. The CVI Questionnaire does not only provide important information about the subjectively experienced visual difficulties. It also allows conclusions to be drawn about possible specific visual problems, which can then be examined more detailed and accurate. The additional examination of cognitive (non-visual) functions also allows a decision as to whether the visual abnormalities are primary or secondary in nature.

The results of our study may be representative for non-clinical service settings specializing in children and teenagers with visual impairments, and therefore cannot be transferred to, for example, a clinical setting, where also younger children with many proven neurological causes are assessed. When comparing the results of our study with those obtained in clinical (e.g., ophthalmological or neuro-ophthalmological) settings, obvious differences can be reported. For example, Fazzi et al. (2007) found a reduction in visual acuity in over 80% of the cases and disturbed eye movements in 30% (saccades) to 80% (following movements) of the cases in their group of 121 children with neurological diagnoses and an average age of 4.5 years at the time of assessment (range: 3 months to 15 years). The outcome in visual-perceptual abilities is not comparable because the authors only reported data on 27 subjects. In the study by Morelli et al. (2022), 51 children with a neurological diagnosis were assessed at an average age of 9 years (range: 5–18 years). Eye movement disorders were identified as ‘core symptom of CVI’, in particular disordered saccadic and smooth pursuit eye movements present at an early stage. In our study, with the exception of the smooth pursuit eye movements, we did not find such high frequencies of oculomotor disorders. Furthermore, we did not find significant differences between the two groups in the type and frequency of oculomotor dysfunction. The different results can probably best be explained by the fact that the subjects reported by Fazzi et al. (2007) and Morelli et al. (2022) suffered more severe oculomotor disorders than the subjects in this present study. Of course, oculomotor dysfunction can indirectly affect visual perception but does not represent primary visual disorders.

In our more natural setting, in most cases, only one examination appointment was possible, and thus only limited examination time was available. In addition, we wanted to keep the mental stress and the fatigue during the examination as low as possible. With the introductory talk beforehand

and the consultation afterwards, the individual appointment lasted between 2.5 and 3 h. The 'price' for this is that in our setting (and probably also in similar settings) only a defined number of tests could be carried out. In individual cases, a second testing session is certainly necessary to gain a more detailed visual and cognitive profile. This seems indicated in cases with visual recognition disorders (developmental visual agnosia), or severe impairment in vision and/or cognition, as, for example, in preterm and term hypoxic children, or in severe developmental delay, with functional disorders in the various cognitive domains (Bathelt et al., 2018; Das et al., 2007; Martin et al., 2016; Tadic et al., 2009).

Our study has several limitations. We were unable to characterize our subjects with regard to brain dysfunction, because information, in particular on critical events during childbirth and on brain imaging, was not available in all cases. Ideally, all participants with CVI+ should present clear evidence of central visual system damage, while in the CVI− group there is no evidence of central visual system damage. However, the presence of a suspected medical diagnosis alone does not seem to be sufficient evidence for the presence of a visual perception disorder, because subjects may suffer from brain dysfunction and nevertheless can have normal visual abilities. Evidence or suspicion for brain dysfunction does not necessarily imply that vision is also impaired. Thus, the presence of CVI can probably better be verified by means of a suitable questionnaire and appropriate visual diagnostic procedures. Setting the total score of the questionnaire to $\geqslant 2$ was somewhat arbitrary. However, we assumed that two visual abnormalities in behaviour are more than random and therefore represent a useful selection criterion. However, it remains to be clarified which items of the questionnaire are the most meaningful in order to consider an investigation to be indicated.

We did not use a developmental factor (e.g., verbal IQ) for matching the two groups because we placed the focus of our study to the visual perception profile. In addition, CVI may be associated with delayed language development (Morelli et al., 2022) and therefore interact with a chosen developmental factor. Furthermore, such a test would have significantly increased the examination time. For a comprehensive assessment of the development, this would certainly be a helpful addition, especially with regard to the development of the vocabulary for naming in the visual modality.

Irrespective of these limitations, we believe that the diagnosis of CVI can be adequately ascertained with our diagnostic approach, also considering that clinical proof of the underlying cause is often difficult and imaging methods are not always conclusive (Boot et al., 2010).

Considering the fact that, on the one hand, not every CNS dysfunction is generally associated with CVI, and on the other hand, in many cases, there are complex visual disturbances without a sufficiently evident CNS cause, the question arises as to whether CVI can be used as a reasonable diagnosis. In cases with a detectable affection of the central visual system and resulting loss of visual function, the diagnosis of CVI is certainly correct. In cases with a detectable brain disorder without specific evidence of damage to the central visual system, CVI can be used as a suspected diagnosis (CVI 'at risk'; Williams et al., 2021), if the existing visual-perceptual disturbances cannot be entirely explained by a peripheral visual system dysfunction, oculomotor abnormalities or cognitive abnormalities. There remains the difficult third group, which has no empirical evidence for brain dysfunction and whose visual perception difficulties cannot be traced back to peripheral causes or cognitive abnormalities. The diagnostic use of CVI is further complicated by the fact that many children have peripheral ophthalmologic abnormalities in addition to the central nervous system aetiology (e.g., Fazzi et al., 2007; Morelli et al., 2022).

We, therefore, propose a more functionally oriented approach based on subjective information on visual difficulties, a detailed visual-perceptual profile and the outcome of the ophthalmological examination. According to the results of this study, this visual profile should routinely include overview/visual search, spatial vision, form and figure perception, and reading (text processing).

Testing of object and face perception and recognition and of topographical orientation is recommended in cases with appropriate hints form everyday behaviour. Defined inclusion and exclusion criteria can help to distinguish primary from secondary visual perception difficulties and to differentiate between peripheral and central visual disturbances. Such an approach can prevent visually disabled children without ophthalmological abnormalities from being classified as 'visually normal' and therefore not receive any treatment or support options (Williams et al., 2021). We agree with McConnell et al. (2020, p. 224) that the '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'. Our study may be able to contribute to this important and necessary process. The early diagnosis and the valid assessment of the positive and negative visual and cognitive performance profiles are crucial prerequisites for tailored measures needed to enhance the cognitive, social and personal development of children with CVI (Chokron et al., 2021; Fazzi et al., 2007; Morelli et al., 2022).

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