

SCOPING REVIEW

Interventions for children with cerebral visual impairment: A scoping review

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Abstract

Aim: To examine the nature and scope of the literature on interventions implemented for children with cerebral visual impairment (CVI).

Method: Using a scoping review methodology, a systematic search of the literature was conducted using four databases including PubMed, Web of Science, Scopus, and Embase. Titles and abstracts were screened and data were extracted and synthesized from full-text, eligible studies. Twenty-three articles were identified and evaluated using quality criteria.

Results: Twenty-three of the 895 studies were included with children aged between 1 month and 17 years 6 months. Interventions were grouped into six categories including visual stimulation, task/environmental adaptations, vision training, acupuncture, stem cell transplantation, and transcranial electric stimulation. Outcome measures examined changes in visual function, functional vision, and visual processing.

Interpretation: Intervention research for children with CVI is in its infancy. Interventions for children with CVI may be beneficial; however, comparing results is difficult due to inconsistency in outcome measures. Most of the intervention studies had lower-level evidence and included small sample sizes. High-quality studies with larger samples and comparison groups are needed to fully understand which evidence-based interventions are most effective for children with CVI. With the incidence of CVI increasing, further development and validation of intervention methods is imperative.

Cerebral visual impairment (CVI) is defined as vision loss that results from damage to the postgeniculate visual pathways.¹ More recently, it has been defined as a confirmable visual dysfunction that cannot be attributed to disorders of the anterior visual pathways or any potentially co-occurring ocular impairment.² CVI is the leading cause of visual impairment in children aged 0 to 3 years in the USA.³ A recent prevalence study estimated that 31.5% of mainstreamed students had one CVI-related vision problem whereas 10.5%

of students had multiple issues.⁴ Comorbid diagnoses commonly coexist with CVI, including cerebral palsy (CP), preterm birth, periventricular leukomalacia, and optic nerve atrophy.⁵⁻⁷ The complexities and deficits associated with CVI warrant a comprehensive evaluation and treatment approach provided by a multidisciplinary team consisting of ophthalmologists, neurologists, optometrists, occupational therapists, and teachers of students with visual impairments, among others.⁵ Visual impairment can negatively impact all aspects of life, including learning and development. However, children with CVI may benefit from interventions to improve functional limitations and overall participation in valued activities. Current practice standards recommend

Abbreviations: CVI, cerebral visual impairment; GAS, Goal Attainment Scale; PEDI-NL-CVI, Pediatric Evaluation of Disability Inventory-CVI Dutch version; RCT, randomized controlled trial; VEP, visual evoked potential.

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implementing early intervention for children with CVI to maximize visual development and functional outcomes.⁸ Many review articles have discussed the importance of and need for evidence-based interventions for children with CVI. However, there are few guidelines for the implementation of interventions into practice.

Systematic and scoping reviews involving children with CVI have been published but are primarily geared toward motor skills and/or visual assessment rather than interventions to improve vision (e.g. visual acuity, contrast sensitivity), functional vision (e.g. visual attention), or functional skills (e.g. dressing, school performance, navigating stairs).^{9,10} Although children with CVI often experience comorbid impairments (e.g. motor), currently available reviews leave many questions regarding visual rehabilitation unanswered. Since CVI can occur with or without the presence of CP, it is essential to consider visual rehabilitative interventions separate from comorbidities and coexisting diagnoses. Other systematic and scoping reviews have focused on assessment and/or diagnostic techniques to evaluate CVI¹⁰ or have discussed CVI interventions as a secondary focus, merely scratching the surface on rehabilitation methods.¹¹ Current research demonstrates a foundation for the evaluation and diagnosis of pediatric CVI with an emphasis to further research to understand the effectiveness of intervention methods. The objective of this scoping review was to examine the nature and scope of the literature on interventions implemented for children with CVI.

METHOD

Protocol and registration

A multidisciplinary team was formed to conduct this scoping review using the frameworks by Arksey and O'Malley¹² and Levac et al.¹³ A protocol was established using the Open Science Framework.¹⁴ The research team agreed on the following five steps: (1) identify the research question; (2) identify relevant studies; (3) select studies; (4) chart the data; and (5) collate, summarize, and report the findings.

Identification of the research question

The following research question was developed to guide the scoping review process: In pediatric patients diagnosed with congenital CVI between the ages of 0 years and 18 years, what interventions have been implemented to improve outcomes?

Identification of relevant studies

Information sources included PubMed, Web of Science, Scopus, and Embase. Variations of the following search strategy were applied to all databases: cortical visual impairment OR cerebral visual impairment OR brain based visual impairment AND pediatric OR child OR children OR adolescent OR

What this paper adds

- Intervention studies for children with cerebral visual impairment (CVI) are sparse.
- Most intervention studies for children with CVI have low-level evidence with low critical appraisal scores.
- High-quality, controlled intervention studies are needed to guide families and clinicians toward evidence-based practice.

infant OR baby AND treatment OR intervention OR rehab OR rehabilitation OR habilitation OR program OR academic. Search terms were established by two reviewers. The last search was conducted on 20th June 2022. No limitations regarding language or publication year were applied.

Study selection

Study titles and abstracts were screened for eligibility independently by two reviewers based on established inclusion criteria. Peer-reviewed articles with individuals between the ages of 0 years and 18 years (at least 80% pediatric), diagnosed with congenital CVI or describing CVI characteristics/symptoms without a formal diagnosis (brain insult + function vision deficits), and with original data were eligible for inclusion. Systematic reviews were determined eligible for the current scoping review if at least 50% of articles included interventions for children with CVI. Excluded systematic reviews were manually searched to include relevant studies. Specific types of gray literature (conference abstracts and textbooks), literature reviews, and/or with individuals with acute CVI (i.e. not congenital) were excluded. Studies published in languages other than English were translated using Google Translate. An initial screening of the inclusion criteria was performed on a small sample of articles comparing agreement. Inclusion criteria were refined after group discussion and by consensus. No restrictions were applied for study design to provide an accurate representation of research available. Full-text reviews were completed independently for all eligible studies by two reviewers. Any disagreements between the two reviewers were resolved by a third reviewer. Reviewers were masked to each other's decisions. References were manually searched and additional intervention studies were reviewed for inclusion.

Charting the data

The full texts of all included articles were read independently by two reviewers. Data extraction was completed independently by two reviewers using the data extraction template in Microsoft Excel. Data extraction fields included: evidence

level; sample population/diagnosis; inclusion/exclusion criteria; study purpose; discipline of the interventionist; intervention type/frequency/duration; assessment procedures; outcome measures; method/design; adverse events; significant results; limitations/gaps; and other study details. Missing data were obtained by contacting the study investigators. All data recorded were confirmed by a third reviewer. Two reviewers performed quality assessments of individual studies independently. The JBI approach to critical appraisal checklists was used to assess the methodological quality of all studies included in the scoping review.¹⁵ One point was granted for 'Yes' responses, 0.5 for 'Unclear,' and 0 for 'No' responses; questions with 'N/A' responses were removed from the calculation for all critical appraisal checklist questions. Studies were assigned a grade of high (>80%), medium (60–80%), or low (<60%) based on the average total point percentages on completion of the critical appraisal checklists. Risk of bias for randomized controlled trial (RCT) studies was assessed using the revised Cochrane risk of bias tool for randomized trials (RoB 2).¹⁶ Additionally, the Oxford Centre for Evidence-Based Medicine was used to assess the methodological quality of each study based on the study design where level I is the highest quality and level V the lowest.¹⁷ Figure S1 shows the quality of each study using both the JBI critical appraisal score and the Oxford Centre for Evidence-Based Medicine levels of evidence.

Collating, summarizing, and reporting findings

Key themes were identified using the extracted data and are described in Table S1. Themes were developed around the intervention type, were discussed by the team, and were presented in the narrative section that follows, describing how the results related to the research question.

RESULTS

Study selection

The initial results of the literature searches yielded 895 articles after the removal of duplicates. Manual searching was completed and six additional studies were identified. After title and abstract screening, 753 articles were excluded because they did not meet the inclusion criteria. Of the 142 studies assessed for eligibility by full-text review, 23 were included from three databases. The most prevalent reasons for exclusion during the full-text review were: (1) did not include CVI intervention (29%); (2) were gray literature (i.e. conference abstract/proceedings, book reviews) (23.5%); and (3) were review articles (13%) (see the PRISMA flow diagram in Figure S2).

Study characteristics

Studies included children diagnosed with congenital CVI or exhibiting CVI behaviors/characteristics without a formal

diagnosis (brain insult + functional vision deficits) aged from 1 month to 17 years 6 months. Study size ranged from $n = 1$ to $n = 388$. The 23 studies included in this scoping review were rated using the Oxford Levels of Evidence and consisted of three studies with level I evidence, one study with level III evidence, eight studies with level IV evidence, and 11 studies with level V evidence, making the overall evidence level low (Table S1 and Figure S1). Themes emerged from the studies included and results were organized by theme including: visual stimulation programs (17%); vision skills training (26%); task adaptations and environmental modifications (30%); stem cell transplantation (13%); acupuncture (9%); and transcranial electric stimulation (5%) (Table S1). The outcome measures examined changes on a standard eye exam (e.g. visual acuity, contrast sensitivity), functional visual skills (e.g. visual attention, visual communication), and communication skills. The overall critical appraisal scores for the studies included were high (>80%) for stem cell transplantation interventions and visual skills training; medium (60–80%) for task adaptations and environmental modifications, acupuncture, and transcranial electrical stimulation; and low (<60%) for visual stimulation programs. Publication years ranged from 1997 to 2021 and 64% were published within the last 10 years. Tables S1 and 1 outline the design, participant, intervention characteristics, and critical appraisal scores of the intervention studies included in this scoping review. The risk of bias assessment results for the RCTs are displayed in Table 2.

Interventions

Visual stimulation programs

Four studies included in this review used visual stimulation programs to improve functional vision and/or basic vision skills. Two were observational studies that examined the efficacy of visual stimulation programs in improving functional vision (e.g. visual attention, visual communication) and standard eye exam measures (e.g. visual acuity, contrast sensitivity) with children diagnosed with perinatal brain damage ($n = 99$; $n = 30$). Visual stimulations consisted of stimulation with everyday materials, bright colors and high-contrast material, materials under ultraviolet light, or lights and lighting material. The interventions were provided daily by the participants' parents for 2 to 18 months; the length of the intervention sessions was not reported. In both studies, significant improvements were observed postintervention in visual attention ($p = 0.001$; $p < 0.001$) and visual communication ($p = 0.001$; $p < 0.001$) skills based on outcome data (e.g. Teller acuity, informal observations).^{18,19} A retrospective, quasi-experimental study of 21 children with CVI who received an intensive visual stimulation program (daily for 4–15 months) performed by their parents demonstrated similar results.²⁰ Visual stimulation involved programs for light reflex (1 minute 30 times per day), outline perception (line, edge, and light detection) for 1 minute, 10 times daily,

TABLE 1 JBI critical appraisal checklists for the studies included in this scoping review

Items	Critical appraisal score													
	Q1	Q2	Q3	Q4	Q5	Q6	Q7	Q8	Q9	Q10	Q11	Q12	Q13	
Alimović et al. ^{18a}	Yes	No	No	Unclear	Unclear	No	No	Yes	No	Yes	X	X	X	4/10
Alimović et al. ^{19a}	Unclear	No	No	Unclear	Unclear	Yes	Yes	Yes	No	Yes	X	X	X	5.5/10
Baker-Nobles and Rutherford ²⁹	Unclear	Yes	Yes	Unclear	Yes	Yes	No	Yes	X	X	X	X	X	6/8
Duke et al. ^{27b}	Yes	Yes	Unclear	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	12.5/13
Farrenkopf et al. ²⁶	Yes	Yes	Yes	No	Yes	Yes	Yes	Yes	X	X	X	X	X	7/8
Fazzi et al. ^{28c}	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	X	X	X	X	9/9
Kamali et al. ^{40a}	Yes	No	Yes	Yes	Yes	Yes	No	Yes	X	X	X	X	X	6/8
Lam et al. ^{30a}	Yes	Yes	Yes	No	No	Yes	No	Yes	X	X	X	X	X	5/8
Lanners et al. ^{21a}	No	No	No	Unclear	Unclear	No	Unclear	No	No	No	X	X	X	1.5/10
Li et al. ^{36a}	Yes	Unclear	Yes	Yes	Unclear	Yes	Yes	Yes	X	X	X	X	X	7/8
Little and Dutton ^{31a}	No	No	No	Yes	Unclear	Yes	No	Yes	Yes	Yes	Yes	Yes	Yes	3.5/8
Luan et al. ^{35b}	Yes	Unclear	Yes	No/A	No/A	Unclear	Yes	Yes	Yes	Yes	Yes	Yes	Yes	9.5/11
Lueck et al. ²⁵	Yes	No	Yes	Yes	Yes	Yes	Yes	Yes	X	X	X	X	X	7/8
Macintyre-Beon et al. ^{33a}	Yes	Yes	Yes	Yes	Unclear	No	No	Yes	X	X	X	X	X	5.5/8
Malkowicz et al. ^{20c}	Yes	Yes	Unclear	Yes	No	Yes	No	Yes	Unclear	X	X	X	X	6/9
McDowell and Budd ³²	No	Unclear	No	No	No	No	No	Unclear	No	Yes	X	X	X	2/10
Philip ^{34a}	Yes	Yes	Yes	Yes	No	No	No	No	X	X	X	X	X	4/8
Salavati et al. ^{22a}	Yes	No	Unclear	Unclear	Unclear	Yes	Yes	Yes	No	Yes	X	X	X	6.5/10
Saviola et al. ^{23a}	Yes	Yes	Yes	Yes	Yes	Yes	Yes	Yes	X	X	X	X	X	8/8
Shroff & Das ^{37a}	Yes	Yes	Yes	Yes	No	Yes	Unclear	Yes	Yes	Yes	X	X	X	8.5/10
Walker and Wegner ^{24a}	Yes	No	Yes	Yes	Yes	Yes	No	Yes	X	X	X	X	X	6/8
Wong et al. ^{38a}	No	No	Yes	No	No	Yes	Yes	Yes	Yes	Yes	X	X	X	6/10
Zhao et al. ^{39b}	Yes	Unclear	Yes	Unclear	Unclear	Unclear	Yes	Yes	Unclear	Yes	Yes	Yes	Yes	10.5/13

^aCase report/series ^bRandomized controlled trial ^cQuasi-experimental. The number of questions on the JBI tools vary based on study design. The 'X' in cells indicate end of critical appraisal checklist (no questions for those items). Refer to Aromataris and Munn¹⁵ for information on checklist questions.

TABLE 2 Revised Cochrane risk of bias for randomized controlled trials

	Randomization process	Effect of assignment to intervention	Effect of adhering to intervention	Missing outcome data	Outcome measurement	Selection of the reported result
Duke et al. ²⁷	Low	Low	Low	Low	Low	Low
Luan et al. ³⁵	Low	Low	Low	Low	Low	Low
Zhao et al. ³⁹	Low	Low	Low	Low	Low	Low

Low, low risk of bias. Refer to Sterne et al.¹⁶ for information on domains and scoring interpretation.

outline bits and slides (simple shape detection against a white or black background) 10 times per day, and developing the ability to see detail within a configuration 10 times per day. Sixteen patients demonstrated significant improvement in visual optimization as demonstrated by an increase in their visual competency levels on the developmental profile from level III or lower pre-intervention to level V or higher post-intervention. Comparable results were also recorded in a case series examining the effects of a visual stimulation program (duration of either 1–3 weeks or 1–4 weeks) on visual skills in children with CVI ($n = 76$). Of the participants completing two treatment courses ($n = 30$), 20 had increased visual attention and spontaneous visual curiosity and required less contrast and light during daily interactions compared to baseline.²¹

Vision skills training

Six vision skills training interventions were identified. The primary focus was on enhancing standard eye exam measures, such as grating acuity, contrast sensitivity, and visual fixation and tracking, as well as functional visual skills, such as visual attention, visual communication, and purposeful gaze/reach by means other than visual stimulation. A case series examined the efficacy of an individualized functional training therapy program targeted to improve gross motor function and functional skills in children with CP and CVI ($n = 5$).²² The program was provided at school (physical therapist) and at home (parents) for a total of 18 weeks. Goal Attainment Scale (GAS) scores improved by at least one point for all participants and all scored significantly higher in the functional skills section on the Pediatric Evaluation of Disability Inventory-CVI Dutch version (PEDI-NL-CVI) compared to baseline. The effects of an individualized rehabilitation program to treat CVI was further examined in a case report of a child aged 2 years 6 months with CVI and *KCNQ2* encephalopathy.²³ The program was aimed at improving visual stimulation, pursuit of eye movements and saccades, and increasing eye contact. Outcomes were measured using LEA Grating Acuity, GAS, and Visual Skills Inventory. Improvements were observed in all outcome measures after the intervention. A case study with a child with CVI and CP tested the feasibility of using an eye gaze training program to teach the visual skills needed to access an augmentative and alternative communication device.²⁴ Before the study, the child communicated only through facial expressions and body movements. Results showed that

at the end of the intervention, the child was able to communicate on average 12 utterances in 25-minute sessions.

The effects of visual skills, visual behaviors, and visual environment management training provided at home were examined in a case report of a 14-month-old child with CVI.²⁵ Postintervention data from video recordings demonstrated a 38% increase in visual reaching behaviors and a 65% increase in visual following behaviors. Similarly, a ‘significant’ increase in functional performance levels, based on coded observations, was noted with physical prompts in a case report examining the effects of physical versus verbal prompts on independent drinking skills (visual fixation + purposeful reach toward cup).²⁶ The study outcomes were subjective observations, not standardized assessments.

Task adaptations and environmental modifications

Seven interventions studies were identified, which used strategies to increase visual function and maximize current functional abilities through adaptations to daily tasks and modifications to the environment. One RCT examined if giving caregivers visual support strategies from the Insight Question Inventory improved the quality of life of children with CP in the intervention group compared to those in the control group who did not receive these strategies.²⁷ The intervention group was asked to use the strategies at least three times per day for 6 weeks. Overall, there was no change in quality of life on the PedsQL 4.0 Generic or PedsQL 3.0 for CP. However, the treatment group showed better quality of life on the subdomains of speech and communication ($p = 0.006$). A non-randomized clinical trial evaluated a 6-month intervention using environmental adaptation and early visual training with infants in the treatment group including infants with CVI ($n = 15$) and peripheral visual impairment ($n = 15$) and used a comparison group consisting of 30 matched infants.²⁸ The visual training was completed at least three times per week for 45-minute sessions for 6 months with parents present and engaged during the sessions. Results indicated that both treatment and comparison groups improved on visual function (e.g. visual acuity and contrast sensitivity; $p < 0.01$). The treatment group significantly improved on all ocular motor functions (visual fixation, smooth pursuit, and reactive saccades; $p < 0.01$), whereas the control group did not. When examining the group with CVI compared to the group with peripheral visual impairment, the subgroup with CVI showed more

improvement in visual acuity compared to the subgroup with peripheral visual impairment ($p < 0.001$). A single-individual case report analyzed the effects of an occupational therapy intervention (once per week for 18 months), using environmental modifications and visual skills training, to maximize residual vision in a 14-month-old with CVI.²⁹ After the intervention, most improvements were observed with environmental modifications, such as high-contrast colored lighting, elimination of clutter, and tactile integration of visual input, and recognized via goal attainment. Similar results were found in another case report describing the prognosis for higher cognitive visual functions in one child with severe CVI over 6 years.³⁰ The intervention consisted of environmental modifications (e.g. simplification) and adaptations to daily activities by using intact alternative sensory pathways to maximize current visual function. Improvements were observed in visual acuity only, with no improvements in higher-level visual skills, based on parent reports and observations. No functional outcomes were reported, although results indicated improved quality of life and encouraged typical development.

The effect of color on visual awareness and attention was examined in a case series using colored tents (a sheet of fabric draped over the child's head) with children with CP and CVI ($n = 2$) in the school setting.³¹ There was no information reported on the duration and frequency of the intervention. However, researchers stated the tents increased visual awareness and attention based on observations during sensory sessions. McDowell and Budd³² presented a case series based on the perspectives of teachers/paraeducators. The study examined whether decluttering the classroom alleviated visual problems and associated behavioral challenges in students with CVI. Classroom changes involved removing unnecessary equipment and furniture, covering glass with black paper, and hanging sheets over open shelving. Teachers/paraeducators observed the students over 2 weeks and reported that the changes had a positive effect on the students' learning experiences, behaviors, and function compared to baseline. However, students continued to be distracted by noise and movements. Details on specific visual problems/behaviors before decluttering were not discussed. A longitudinal case study showed strategies relating to navigating unfamiliar areas and minimizing clutter as being the most helpful in alleviating visual deficits based on a self-reported rating scale.³³ Similarly, strategies involving tactile guide/recognition while navigating and reducing distractions resulted in significant progress in visual abilities for a child with CVI (e.g. increased eye contact and interest in surroundings) in another case report.³⁴

Stem cell transplantation

Three stem cell transplantation intervention studies were identified, using stem cells from participants' bone marrow, or derived from fetus donors/human embryonic cells. In an RCT, the treatment group received intracerebroventricular

neural stem/progenitor cell transplantation ($8 \times 10^6 - 10 \times 10^6$ neural stem/progenitor cells, once in total) and rehabilitation training. The control group received rehabilitation training only. Visual function improved by at least one level on the Hoyt method for children with a normal fundus exam in 64% of the treatment group and 33% of the control group.³⁵ The rate at which visual improvement was noted varied significantly among both groups. One child with an abnormal fundus exam improved in the treatment group whereas none (0 out of 4) with an abnormal fundus exam improved in the control group. A subset of children in both groups received functional magnetic resonance imaging (fMRI). The fMRI was analyzed qualitatively looking for positive and negative signals with mixed results in both groups. The fMRI results between groups showed borderline statistical significance ($p = 0.052$) for participants with periventricular leukomalacia only. Comparable findings were shown in a case report of an 11-year-old child with CP and CVI treated with intravenous bone marrow mesenchymal stem cell transplantation (4×10^7 cells infusions, four times for 20 days).³⁶ Significant improvements in visual function based on visual acuity and visual evoked potential (VEP) testing 1-year post-bone marrow mesenchymal stem cell treatment compared to baseline were noted. These findings are further supported by an observational study of children with CP and CVI ($n = 40$) analyzing the efficacy of human embryonic stem cell therapy.³⁷ After four treatment phases (twice per week, 0.25ml/day human embryonic stem cells), the participants' Nutech functional scores improved by a minimum of one level; single-photon emission computed tomography demonstrated significant improvements in perfusion. No statistically significant differences between participant age, sex, cause of CVI, or brain injury type were reported for all three studies.

Acupuncture

Two acupuncture intervention studies were identified. Wong et al.³⁸ examined the efficacy of tongue and body acupuncture treatment (60 sessions for 6 months) in improving visual recovery in a case series of 12 children with CVI. Clinical and functional improvements (e.g. visual acuity and/or improvement of one level on the Visual Functional Scale) were observed in 33% of participants, improved VEPs in 56%, improved brain positron emission tomography glucose metabolism in 85%, and 58% improvement via the Clinical Global Impression Scale. In those who made clinical or functional improvements, treatment intervals were more than twice as long. No correlations were identified between cause, severity, or clinical/functional visual outcomes with positron emission tomography or VEP improvements. An RCT examined the clinical effectiveness of acupuncture in conjunction with visual stimulation therapy in children with CVI ($n = 60$) aged 3 to 10 years.³⁹ Children in the treatment group ($n = 30$) received visual stimulation twice per day and acupuncture three times per week for 4 weeks as a course of treatment, for three courses in total. Acupuncture was conducted on the

scalp to the visual area and auxiliary points. Each time, four main points and one to two auxiliary points were used. The control group ($n = 30$, visual stimulation only) received three courses of visual stimulation and each course lasted 4 weeks. After treatment, both groups demonstrated improved pattern VEPs with shorter latency period (P100-L) and higher amplitude (P100-A). The amount of change was greater in the treatment group ($p < 0.05$). The treatment group demonstrated more improvements in visual acuity compared to the control group ($p < 0.05$) posttreatment. Although both groups demonstrated improvements in these areas, the treatment group results showed a larger clinical effect.

Transcranial electrical stimulation

In a case study, an 11-year-old with CVI participated concurrently in visual rehabilitation therapy, transcranial electrical stimulation, and pharmacological therapy. They received medication, participated in a self-training eye exercise (Fit Eye) daily for 30 minutes, twice per day for 2 months.⁴⁰ The self-training eye exercises were completed 40cm from a 15-inch monitor, with 60% luminance; they were done with the dominant hand, in a dark room for 30 minutes and 30 minutes later in a well-lit room, every day for 2 months. They also completed a mirror-tracing task, tracking a spiral maze at different luminance levels and distances for 30 minutes every other day for a 60-day follow-up period. They also received five sessions of transcranial electrical stimulation in the morning followed by five sessions of repetitive transorbital alternating current stimulation in the afternoon for 5 consecutive days. Results showed changes in the brain after the intervention, improvements in visual acuity, and improvements on the Low Vision Quality-of-Life Questionnaire.

DISCUSSION

The objective of this scoping review was to examine the nature and scope of the literature available on interventions implemented for children with CVI. Unlike other reviews, this scoping review encompassed a broad perspective including all intervention studies regardless of discipline, provider, or study design. This review excluded medical and/or surgical interventions used to treat medical conditions in children with or at high risk for CVI since treatment was directed toward the medical condition not the functional outcomes (e.g. cooling intervention for infants with hypoxic-ischemic encephalopathy and/or hemispherectomy for seizure control).

Quality of evidence

Our review included three RCTs, which had a high level of evidence; all three had high critical appraisal scores. These RCTs varied in the interventions delivered and included task

adaptations and environmental modifications, stem cell transplantation, and acupuncture. However, the majority of the studies in this review had low-level evidence (levels IV and V) with medium-to-low critical appraisal scores. There were six studies in the review with low-level evidence and high critical appraisal. The interventions studied in this scoping review may have potential in the future, but these studies are difficult to generalize without additional research using more rigorous study designs.

Intervention outcomes

The studies included assessed intervention effectiveness using either standard eye exam measurements (e.g. visual acuity, eye alignment), functional vision skills (e.g. visual attention, visual communication), neurophysiological measures (e.g. VEP, fMRI), or functional skills (e.g. GAS, PEDI-NL-CVI, communication). The most frequently used outcome measures were those used during a standard eye exam (visual acuity and contrast sensitivity), which evaluate the anterior visual pathway. This is supported by Swaminathan and Deiva Jayaraman¹¹ who stated that quantitative tests, such as Teller acuity cards, are crucial in establishing a baseline to monitor improvement in children with CVI. Although visual acuity tests have been validated in many populations and have high test-retest reliability, these tests have not yet been studied to understand their correlation with improvement in higher-order visual processing that assesses post-geniculate visual pathway function. Furthermore, studies in this review lacked outcome measures that assess higher-order visual processing. Three RCTs and one non-RCT reported statistically significant improvements in outcomes compared to controls. These studies also had higher critical appraisal scores. Two studies implemented task adaptations and environmental modifications, one implemented stem cell transplantation, and one acupuncture. However, these studies cannot be compared because they all had different outcome measures making comparison impossible.

Eleven studies used measures of functional vision skills. However, most of these measures were subjective, thereby increasing the risk of bias, and/or were non-standardized, creating issues with reproducibility, indicating weak generalizability and field application. Furthermore, it is unclear whether subjective measures were used to evaluate functional vision due to clinician preference, lack of available valid and reliable functional vision outcome measures, or inability to use standardized eye exam measures.

Although the intervention studies included reported some degree of improvement for children with CVI, the overall level of evidence was low to medium, minimizing reliability, validity, and generalization of results. An essential conclusion to highlight is the immense lack of functional outcomes and quantitative, objective outcome measures used in the included intervention studies. Research using more quantitative assessment of functional vision and functional outcome measures is needed.

Interventions

Based on the Oxford Levels of Evidence and the JBI critical appraisal scores, there is no intervention that stands out as being a high-quality, evidence-based intervention. When rating the methodological quality of a study, many factors are considered including study design, study quality, masking of assessors, use of validated and reliable outcome measures, and reporting of the results. Only a few studies in this review had high-quality evidence on the Oxford Levels of Evidence and high critical appraisal scores. Most of the studies included had lower quality of evidence and small sample sizes making generalizability of results impossible; therefore, interventions are weakly recommended without further controlled testing.

It is difficult to decipher if the improvements observed after intervention in children with CVI are a result of natural, expected processes due to age/developmental phase or from the intervention itself, which can lead to ambiguity and should be considered. The plethora of confounding variables impacting the effects of interventions for children with CVI makes it challenging to ascertain accurate correlations between interventions and outcomes. This finding is supported through a common theme recognized in the task and environmental adaptation intervention studies. Researchers concluded that visual deficits are more difficult to alleviate with the existence of pervasive visual dysfunction. Many of the studies lacked specificity regarding the severity of CVI or had inconsistent (e.g. non-standardized) methods in recognizing CVI severity, making conclusions difficult to support without accounting for such variables. Most of the studies included in this scoping review involved children with a confirmed diagnosis of CVI but etiology was not consistently addressed. Since CVI varies in complexity, accounting for all confounding variables is vital. However, such intricacies may be the reason why few formal, rigorous studies have been completed. Additionally, future studies should consider incorporating control groups or multiple baseline designs.

Two studies used functional outcome measures to examine changes in participant goals and motor skills (GAS and PEDI-NL-CVI). The GAS measures changes in child- and family-identified goals and the PEDI-NL-CVI measures changes in functional skills in children with CP and CVI (e.g. self-care, mobility). The use of functional outcome measures to bridge the gap from improving functional vision (e.g. the use of vision during daily activities) to improving functional performance is essential. All future studies should include at least one functional outcome measure, making the link between vision and function imperative.

Limitations

One study limitation is a risk of publication bias and misrepresentation due to the limited amount of evidence available. Furthermore, most of the studies included in this

scoping review did not use masked assessors and rarely used the same outcome measure, making it difficult to compare study findings. The complexity of CVI may negatively impact intervention results and decrease the likelihood of findings being published. Further risk of bias exists because of certain types of gray literature (conference abstracts and book reviews) were excluded.

Future research

The current body of evidence describing interventions for children with CVI is limited, with low levels of evidence. Previous reviews focused on assessment or diagnostic methods for CVI rather than interventions. The findings of this review demonstrate that there is insufficient evidence to guide clinicians toward providing evidence-based interventions. The need for advancements in CVI intervention methods has been recognized and suggested for nearly two decades.⁴¹ However, the results of this review indicate that research efforts are scarce; many studies were older, with lower-level evidence.

Furthermore, there is a strong need for assessments of functional vision that are valid, reliable, and can be used as outcome measures before and after visual interventions.^{42,43} Future research should evaluate the correlation between assessments for anterior and postgeniculate visual pathways to discover if they can be used interchangeably and development and implementation of reliable functional assessments. Additionally, higher-quality, controlled research studies are needed due to the substantial rate of spontaneous improvement reported in previous papers.

Conclusions

Although there are few published interventions available for children diagnosed with CVI, there are no interventions for CVI with efficacy demonstrated by high-quality, controlled studies. Many studies used subjective measures to assess intervention effectiveness. Although the interventions found in this scoping review showed improvements in standard eye exam measures and/or functional vision, the overall level of evidence is low due to small sample sizes, lower-level study design, and/or lack of comparison groups. With increased survival rates of children born preterm and with complex medical conditions, the incidence of CVI continues to rise; thus, further investigation to develop and validate intervention methods is imperative. Although there is a lack of large-scale RCTs for pediatric CVI interventions, the potential exists for future, larger controlled trials of interventions identified in this review. Future studies should use objective outcome measures to monitor improvements and analyze confounding variables. Additional research is warranted to establish best practice for children diagnosed with CVI.

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DATA AVAILABILITY STATEMENT

Data sharing not applicable - no new data generated, or the article describes entirely theoretical research.

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SUPPORTING INFORMATION

The following additional material may be found online:

Figure S1: Methodological quality.

Figure S2: Flow diagram for the studies included in this scoping review.

Table S1: Thematic analysis of interventions.

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