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A Systematic Review on the Management of Cortical Visual Impairment

Myunghwa Oh[a], **Hyunjoong Kim**[b*]

[a]Department of Occupational Therapy, Dongshin University, Republic of Korea
[b]Department of Senior Exercise Prescription, Gwangju Health University, Republic of Korea

Objective: Cortical Visual Impairment (CVI) is a leading cause of pediatric blindness and the most common form of pediatric visual disability, particularly prevalent among children with cerebral palsy (CP). This systematic review synthesizes the latest research on various interventions for managing CVI, focusing on studies published in the last decade.

Design: A systematic review

Methods: A comprehensive search was conducted in March 2024 across several databases including MEDLINE, CINAHL, Embase, and Web of Science. Studies were selected based on inclusion criteria set under the PICOSD framework and were limited to those involving human subjects, published in English, and conducted within the past ten years. The selected studies included randomized controlled trials, observational studies, and case reports focusing on rehabilitation, therapy, and surgical interventions for CVI.

Results: Out of 221 studies screened, 5 met the inclusion criteria and were reviewed in detail. These studies covered a range of interventions including physiotherapy, sensory integration training, visual training programs, neuromotor rehabilitation, and surgical procedures aimed at improving visual function and overall quality of life for CVI patients.

Conclusions: The studies demonstrate the potential benefits of structured, early intervention programs that incorporate family involvement and are tailored to the unique needs of children with CVI. However, there remains a significant need for further research to establish evidence-based practices in this field.

Key Words: Visual impairments, Cerebral palsy, Neurorehabilitation

Introduction

Cortical Visual Impairment (CVI) is one of the leading causes of pediatric blindness [1] and is the most common type of pediatric visual impairment [2]. Notably, up to 83% of individuals with cerebral palsy also have CVI, as neurological damage affecting the visual pathways can also impact the cortical spinal tract; thus, these disorders often co-occur [3, 4]. Diagnostic studies have reported a correlation between bilateral spastic cerebral palsy and CVI [5].

The causes of CVI typically originate from brain damage at birth, with contributing factors including hypoxia, ischemia, head injury or trauma, infections such as encephalitis or meningitis, seizure disorders, genetic disorders, and metabolic disorders [6, 7]. Radiologically, CVI is characterized by significantly underdeveloped major white matter areas related to visual processing [8]. Most importantly, given that CVI leads to impaired vision, there is a critical need for proactive management during the first two years of life to mitigate its impact on normal development through neuroplasticity, as this is a period when visual developmental stimuli are limited [9, 10].

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Corresponding author: Hyunjoong Kim (**ORCID** https://orcid.org/0000-0001-6538-3872)
Department of Senior Exercise Prescription, Gwangju Health University
73 Bungmun-daero 419beon-gil, Sinchang-dong, Gwangsan-gu, Gwangju [62287], Republic of Korea
Tel: ***-****-**** **Fax:** +82-62-958-7610 **E-mail:** hjkim@ghu.ac.kr

General management of CVI includes training in vision, physical, cognitive, and behavioral skills [11-13]. Managing CVI in children involves early diagnosis to assess residual vision and aim for the minimization of disability to enhance quality of life [14]. Children with CVI are not generally affected in terms of mortality but often live with visual impairments for life, necessitating opportunities for visual function and social participation [15].

Despite the acknowledged importance of early intervention in the management of CVI, both domestically and internationally, there is a lack of research and no consensus on evidence-based treatments reported so far. Therefore, this review aims to synthesize and analyze studies on the management of CVI from the past decade.

Methods

Study Design

This study is a systematic review that synthesizes research on the management of CVI and conducts a qualitative analysis. This review was conducted in accordance with the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) guidelines.

Eligibility Criteria

The eligibility criteria were structured according to the key question strategy PICOSD (Participants [P], Intervention [I], Comparison [C], Outcomes [O], Study Design [SD]). The selection criteria included participants with cerebral palsy and CVI, and the interventions were set as rehabilitation, exercise, therapy, training, and comprehensive management strategies. Since only a qualitative analysis of the management was included, no comparison group was set. Outcome variables included all evaluation tools to monitor the progression of symptoms of CVI. The study design of this review included intervention studies. Studies not involving human subjects, studies not written in English, research reported at academic conferences, and studies older than 10 years were excluded.

Search Strategy

In this review, the searches were independently conducted in March 2024 by two researchers experienced in meta-analysis. The search strategy combined terms representing the P and I and was conducted with reference to medical subject headings (MeSH). The search included pre-identified keywords (cerebral palsy) AND (cerebral visual impairment OR CVI OR cortical visual impairment OR cognitive visual dysfunction OR visuoperceptual disorder OR visual disorders OR higher visual disorder) AND (rehabilitation OR care OR therapy OR disease management'/exp OR treatment OR exercise OR neurorehabilitation OR training) and index terms, utilizing international electronic databases such as the Cumulative Index of Nursing and Allied Health Literature (CINAHL), Excerpta Medica Database (Embase), Medical Literature Analysis and Retrieval System Online (MEDLINE), and Web of Science.

Data Extraction

Studies identified from the aforementioned electronic databases were extracted and duplicates were removed using Microsoft Excel (Microsoft, Redmond, Washington, USA). Following the PRISMA guidelines, each study's title and abstract were reviewed before proceeding to full-text review. If there were any discrepancies, the full texts were reviewed together to finalize the selection.

Risk of Bias Assessment

For randomized controlled trials, a risk of bias (RoB) assessment tool consisting of seven items developed by the Cochrane Bias Methods Group was used [16]. To evaluate the quality of the studies, two researchers assessed the risk of bias as low (+), high (-), or unclear (?). Items that were not in agreement were resolved by jointly reviewing the original articles to reach a consensus. Cross-sectional studies used the AXIS (Appraisal tool for Cross-Sectional Studies) tool [17], and the risk of bias was evaluated as present (+), absent (-), or unclear/irrelevant (?). Case reports were assessed using the CACR (Critical Appraisal Checklist for Case Reports) [18], and the risk of bias

was evaluated as present, absent, or unclear.

Results

Literature Search and Characteristics of the Included Studies

Out of a total of 221 studies identified through international databases, 66 were checked in Excel to remove duplicates. Following the first screening based on titles and abstracts, 149 studies were excluded, and an additional study was excluded due to inappropriate research design after full-text review, totaling 150 exclusions. Ultimately, 216 studies were excluded, and 5 studies were finally included for qualitative analysis [19-23] (Figure 1).

Risk of Bias Assessment for Enrolled Studies

The randomized controlled trial accounted for one study [19] with the following RoB results: random sequence generation (+), allocation concealment (+), blinding of participants and personnel (−), blinding of outcome assessment (−), incomplete outcome data (+), risk of selective reporting (?), and other biases (+). Observational studies were evaluated using AXIS for three studies [20, 22, 23], and the results are shown in Table 1. Additionally, one case report [21] was assessed using CACR, and the results are in Table 2.

Management of cortical visual impairment

In recent studies, a variety of tailored interventions have been developed to address the unique challenges faced by children with CVI. Cemali et al., 2022 [19], integrated standard physiotherapy with sensory integration training in their approach, providing two 45-minute sessions weekly over an eight-week period to enhance physical functions. Fazzi et al., 2021 [20], introduced a visual training program that features early

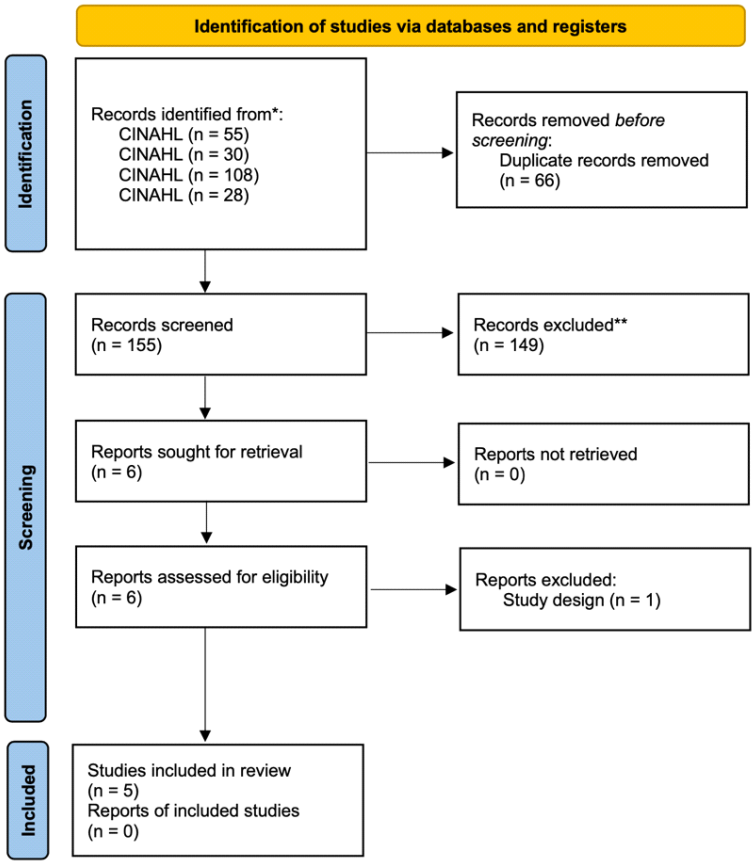

Figure 1. PRISMA flow diagram

Table 1. Appraisal tool for Cross-Sectional Studies.

Contents	Questions	Fazzi, et al., 2021 [20]	West, et al., 2021 [22]	Ye, et al., 2022 [23]
Introduction	Were the aims/objectives of the study clear?	+	+	+
Methods	Was the study design appropriate for the stated aim(s)?	+	+	+
	Was the sample size justified?	−	−	−
	Was the target/reference population clearly defined? (Is it clear who the research was about?)	+	+	+
	Was the sample frame taken from an appropriate population base so that it closely represented the target/reference population under investigation?	+	+	+
	Was the selection process likely to select subjects/participants that were representative of the target/reference population under investigation?	+	−	−
	Were measures undertaken to address and categorise non-responders?		−	−
	Were the risk factor and outcome variables measured appropriate to the aims of the study?	+	+	+
	Were the risk factor and outcome variables measured correctly using instruments/measurements that had been trialled, piloted or published previously?	+	+	+
	Is it clear what was used to determined statistical significance and/or precision estimates? (eg, p values, CIs)	+	+	+
	Were the methods (including statistical methods) sufficiently described to enable them to be repeated?	+	+	+
Results	Were the basic data adequately described?	+	+	+
	Does the response rate raise concerns about non-response bias?	+	−	−
	If appropriate, was information about non-responders described?	+	−	−
	Were the results internally consistent?	+	+	+
	Were the results for the analyses described in the methods, presented?	+	+	+
Discussion	Were the authors' discussions and conclusions justified by the results?	+	+	+
	Were the limitations of the study discussed?	+	+	+
Other	Were there any funding sources or conflicts of interest that may affect the authors' interpretation of the results?	+	+	+
	Was ethical approval or consent of participants attained?	+	−	+

intervention for participants aged between 4 and 12 months corrected age, delivering at least three 45-minute sessions weekly for six months, and incorporated a family-oriented approach by involving parents in the sessions and the broader rehabilitation program.

Saviola et al., 2018 [21], outlined a neuromotor rehabilitation plan that included physiotherapy sessions twice a week for six months, aiming to increase proximal strength and enhance axial stability through

Table 2. Critical Appraisal Checklist for Case Reports.

Questions	Saviola, et al., 2018 [21]		
	Yes	No	Uncertain
Were patient's demographic characteristics clearly described?	✓		
Was the patient's history clearly described and presented as a timeline?	✓		
Was the current clinical condition of the patient on presentation clearly described?	✓		
Were diagnostic tests or assessment methods and the results clearly described?	✓		
Was the intervention(s) or treatment procedure(s) clearly described?			✓
Was the post-intervention clinical condition clearly described?	✓		
Were adverse events (harms) or unanticipated events identified and described?		✓	
Does the case report provide takeaway lessons?			✓

exercises in various positions such as supine, prone, sitting, and transitioning from lying to sitting. West et al., 2021 [22], reviewed medical records to track the postoperative progress of children with CVI, with and without CP, emphasizing the importance of comprehensive monitoring of recovery and adjustments post-surgery. Additionally, Ye et al., 2022 [23], detailed an approach involving microsurgery of bilateral lateral rectus muscle recession in children with exotropia, CP, and CVI, which aimed to correct visual alignment issues directly related to these conditions.

Each of these interventions demonstrates a specific approach, combining medical, surgical, and therapeutic strategies to optimize outcomes for children with CVI (Table 3).

Discussion

The management of CVI in children necessitates a multifaceted approach that addresses the complex interplay between visual impairment and neurodevelopmental needs. This systematic review highlighted several key interventions that have been explored in recent years, each aiming to address specific aspects of CVI management.

The integration of sensory and motor training, as demonstrated by Cemali et al., 2022 [19], forms a foundational therapeutic approach, highlighting that regular, intensive sessions can significantly influence physical development and sensory integration. This is crucial for managing CVI, where enhanced muscle strength and stability support the physical needs of affected children, who often face multiple physical challenges.

Fazzi et al., 2021 [20], stress the importance of early visual stimulation, suggesting that initiating visual training programs during critical developmental windows can substantially improve visual processing capabilities. This early intervention is vital in mitigating the impacts of CVI by enhancing the brain's ability to process visual information more effectively. Early intervention in CVI is paramount to harnessing the neuroplastic capabilities of the young brain, which can significantly improve the prognosis for visual and neurodevelopmental outcomes [24, 25]. Starting treatment during key developmental windows enables more effective neural adaptation, helping to mitigate the long-term impacts of visual impairments. Programs focused on early visual and sensory stimulation can promote essential neural pathways development, contributing to better functional vision and overall cognitive development [26-28].

Saviola et al., 2018 [21], propose a neuromotor rehabilitation strategy that concentrates on increasing muscle strength and stability, essential for children with CVI to navigate their environments more effectively. Enhanced muscle strength is crucial for maintaining posture and balance, which are fundamental for independence and mobility in children with physical disabilities [29, 30].

Additionally, Ye et al., 2022 [23], underscore the

Table 3. Characteristics of the trials.

Study	Study Design	Participants (n)	Intervention	Outcomes
Cemali, et al., 2022 [19]	Randomized controlled trial	CP＋CVI EG (17), CG (17)	Both groups were given physiotherapy training as 2 sessions of 45 min per week for 8 weeks. In addition to the physiotherapy training, the intervention group received sensory integration training as 2 sessions of 45 min per week for 8 weeks.	• ensory function: Alberta infant motor scale, test of sensory functions in infants
Fazzi, et al., 2021 [20]	Intervention clinical trial	EG (30), CG (30) CVI (15) PVI (15)	The visual training programme implemented in this study was characterized as early (applied when participants were aged between 4 and 12mo corrected age), intensive (at least three 45-min sessions per week for 6mo), and family-oriented (parents were present during the training sessions and engaged in the rehabilitation programme).	• eurological test: Amiel-Tision protocol • isual function: cycloplegic refraction, anterior segment, and ocular fundus examination, visual acuity and contrast sensitivity, fixation, smooth pursuit, and reactive saccades • evelopmental skills: GMDS
Saviola, et al., 2018 [21]	Case report	CVI (1)	The neuromotor rehabilitation plan for the patient included physiotherapy twice a week for 6 months (setting: supine, prone, sitting with and without anterior table, on side, long-sitting, transition from supine to sitting by rolling on sides) aimed at increasing proximal, girdle and trunk musculature and improving axial stability.	LEA grating acuity test, goal attainment scale, visual skills inventory
West, et al., 2021 [22]	Retrospective cohort study	CP＋CVI (151) CVI (153)	Medical records of the postoperative course of all children with CVI, with and without CP, were reviewed.	visual acuity refractive error, ocular alignment: alternate cover prism test(or Krimsky test) optic atrophy congenital optic nerve anomaly
Ye, et al., 2022 [23]	Intervention clinical trial	CP＋CVI (38)	Exotropia with CP and CVI children were performed microsurgery of bilateral lateral rectus muscle recession.	PCT CST BCVA

BCVA: best corrected visual acuity, CG: control group, CP: cerebral palsy, CST: contrast sensitivity testing, CVI: cortical visual impairment, EG: experimental group, GMDS: Griffiths mental developmental scales, PCT: prism cover test, PVI: peripheral visual impairment.

significance of surgical intervention in managing CVI, particularly through procedures like bilateral lateral rectus muscle recession. These surgeries correct visual alignment issues, potentially enhancing the visual field

and alleviating the strain caused by CVI [22, 23]. This surgical approach addresses the anatomical and structural problems contributing to visual impairment, thereby not only improving visual function but also reducing physical discomfort associated with CVI [31, 32].

Despite these promising approaches, the review identifies a glaring gap in consensus and standardization across treatment protocols. The limited number of studies, coupled with a lack of long-term follow-up, underscores the need for ongoing research. Additionally, the varying methodologies and outcome measures used in these studies point to the necessity for establishing more uniform frameworks for assessing treatment efficacy.

Continued research should aim to consolidate these diverse strategies into cohesive treatment guidelines that can be universally applied. Further, exploring the integration of technological advancements such as digital aids and virtual reality could offer new avenues for enhancing visual rehabilitation [33, 34]. Establishing a global consensus on CVI management will not only improve treatment outcomes but also aid in the development of standardized training protocols for healthcare providers.

Managing CVI in children requires a holistic approach that targets not only the visual impairment but also considers the wide range of developmental issues these children may face. As highlighted in the systematic review, integrating sensory and motor training is crucial. This dual approach can significantly help in the neurodevelopmental growth of children suffering from CVI, enhancing their ability to process sensory information and respond more effectively to their environment. Early visual stimulation, initiated during critical developmental periods, can significantly enhance the brain's ability to process visual inputs [35, 36]. This is particularly important for optimizing visual pathways that are not fully developed or are damaged. Using structured visual training programs can accelerate visual development and enhance cognitive visual functions.

This review advocates for an evidence-based approach to the management of CVI, emphasizing the importance of early intervention and the potential for integrating various therapeutic modalities to optimize outcomes for this vulnerable population.

Conclusion

This systematic review highlights the diverse approaches to managing CVI in children, emphasizing the need for early and multifaceted interventions. Despite the promising potential of these interventions, the review reveals a significant gap in standardized treatment protocols and long-term effectiveness data. Future research should focus on developing evidence-based guidelines and exploring innovative technologies to improve outcomes for children with CVI. Enhanced collaboration and research are essential to establish effective, standardized care that can adapt to the unique needs of each patient.

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Conflict of interest

The authors of this study declare that there were no potential conflicts of interest with respect to the research, authorship, and publication.

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