RESEARCH ARTICLE

Clinical Manifestations of Cortical Visual Impairment in Department of Ophthalmology, Dr. Cipto Mangunkusumo National Hospital

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Abstract

Cortical visual impairment (CVI) is one of the primary causes of visual function disorders in children under the age of five years. It requires multi-disciplinary involvement due to its various causes, manifestations, and treatments. However, there are no specific guidelines for detecting and diagnosing CVI. The objective of this study is to provide an overview of clinical characteristics and related medical histories for the diagnosis of CVI. This is a cross-sectional study conducted based on medical records at Cipto Mangunkusumo National Hospital Kirana. Data collection was carried out from January 1 to December 31, 2016. The study employed all the children with CVI who were not accompanied by additional visual abnormalities other than nystagmus and strabismus. The analysis was carried out using the Statistical Package for Social Sciences (SPSS) software. There were 16 patients who meet the inclusion criteria. The history of seizures and retardation of growth and development was experienced respectively 14/16 and 13/16 patients. Intracranial infections and epilepsy were the most common comorbidities. Strabismus is the most common clinical pathology (8/16) and 13/16 patients could not pass the light and object fixation test. Flash visual-evoked-potential (VEP) examination revealed that 5/8 patients had a decrease in amplitude, and 7/8 patients had an extension on the latency period. The knowledge of clinical characteristics e.g. decreased visual function, strabismus, or nystagmus that was accompanied by a history of seizures, or growth and developmental disorders can help to detect CVI.
Keywords*: cortical visual impairment, epilepsy, nystagmus, strabismus.*

Manifestasi Klinis dari Cortical Visual Impairment di Departemen Oftalmologi RSUPN dr. Cipto Mangunkusumo

Abstrak

Cortical Visual Impairment (CVI) merupakan salah satu penyebab utama gangguan fungsi visual pada anak dibawah usia lima tahun. Berbagai macam etiologi, manifestasi, dan tatalaksana pada CVI menyebabkan perlunya keterlibatan berbagai macam disiplin ilmu. Namun hingga saat ini belum terdapat pedoman khusus untuk mendeteksi dan mendiagnosis CVI. Oleh karena itu, penelitian ini diharapkan dapat memberikan gambaran karakteristik klinik dan riwayat kesehatan yang berhubungan untuk membantu mendiagnosis CVI. Studi dilakukan secara cross-sectional berdasarkan data rekam medis di RSUPN Cipto Mangunkusumo Kirana. Pengumpulan data dilakukan sejak tanggal 1 Januari sampai 31 Desember 2016. Seluruh pasien penderita CVI yang datang tanpa disertai dengan kelainan penglihatan tambahan selain nistagmus atau strabismus menjadi sampel. Analisis dilakukan menggunakan program SPSS 23.00. Terdapat 16 pasien yang memenuhi kriteria inklusi. Riwayat kejang dan gangguan pertumbuhan perkembangan dialami masing-masing 14/16 dan 13/16 pasien. Infeksi intrakranial dan epilepsi menjadi penyakit penyerta yang paling banyak. Gambaran klinis berupa strabismus paling banyak ditemukan (8/16) dan sebanyak 13/16 pasien tidak dapat melakukan fiksasi cahaya dan objek. Pemeriksaan penunjang berupa flash VEP menunjukkan penurunan amplitudo pada 5/8 responden dan perpanjangan masa latensi pada 7/8 penderita CVI. Karakteristik klinis berupa penurunan fungsi penglihatan, strabismus, atau nistagmus yang disertai dengan riwayat kejang atau gangguan tumbuh-kembang dapat membantu mendiagnosis CVI.
Kata kunci*: cortical visual impairment, epilepsy, nistagmus, strabismus.*

Introduction

Cortical visual impairment (CVI) is defined as an impairment of visual acuity involving the posterior visual pathway with minimal or nonexistent anterior visual pathway morbidity.[1-3] A retrospective study in 5 Nordic countries, stated that CVI frequency has increased from 11% to 23%.[4] In addition, The Blind Babies Foundation of Northern California reported that CVI has become a major cause of visual impairment in children under the age of five years.[5]

Various factors can lead to CVI, such as premature birth, hypoxic-ischemic damage, central nervous system infection, stroke, head injury, anomalies in the brain structure, hydrocephalus, chromosome damage, and seizure.[6] These diversities have led to the collaboration of several disciplines (pediatric ophthalmology or pediatric neuro-ophthalmology, pediatric neurology, psychology, and physiotherapy) for the diagnosis and treatment of CVI. Hitherto, the are no specific guidelines available for CVI.[7] However, several approaches can be followed to make the diagnosis. There is a tool for evaluating the patient's neurobehavioral disorders and few ophthalmological examinations to exclude any other eye diseases. In addition, there is electroretinography (ERG), visual evoked potential (VEP), and imaging (ultrasonography/ USG, computerized tomography scanning/CT scan, magnetic resonance imaging/MRI) to confirm the diagnosis.[8] The objective of this study is to provide an overview of the characteristics, clinical features, comorbidities, and workups that can be used to diagnose CVI.

Methods

This study was conducted using a descriptive cross-sectional method. Data regarding age, gender, type of birth, gestational age, birth weight, history of growth and development, history of seizures, comorbidities, results of ERG and VEP, streak results, presence or absence of strabismus and nystagmus, visual acuity, and results of MRI or CT scan of the head were obtained from the medical records of Dr. Cipto Mangunkusumo National Hospital (CMNH) Kirana. Data collection was carried out from January 1 to December 31, 2016. The study employed all the patients with complete medical records and without any other eye disorders except for strabismus or nystagmus during the study.

Data processing was carried out by using the Statistical Package for Social Sciences (SPSS) software version 23. Categorical variables were presented in the form of numbers, whereas numerical variables in the form of mean and standard deviation (for data with normal distribution) or median and range (for data with abnormal distribution).

Result

To simplify the data about maternal age, we grouped the data into "term" and "preterm." Most patients were born through normal delivery (12 subjects), and there was no patient with premature birth. The patient's average age was 20 months (range from 4 months to 60 months old). Retardation of growth and development occurred in the majority of patients (13/16), and only two patients had not had a history of previous seizures. The most common comorbidities were intracranial infections and epilepsy and the other comorbidities that accompanied the diagnosis of CVI are shown in Table 1.

Table 1. Demographic Characteristic of Patients with CVI in the Department of Ophthalmology CMNH Kirana

Demographic characteristics	Frequency (n=16)
Gender	
Boys	11
Girls	5
Birth methods	
Normal	12
Sectio Caesarea	4
Gestational age	
Term	16
Premature	0
Growth and Development	
Normal	3
Delay	13
History of seizure	
Yes	14
No	2
Comorbidities	
Cerebral palsy and epilepsy	3
Cerebral palsy and microcephaly	1
Cerebral palsy	1
Epilepsy	2
Hydrocephalus	1
Microcephaly	1
Intracranial bleeding	1
Infection of central nervous system	4
Cerebral atrophy	1
Febrile seizures	1

Clinical examination revealed that eight patients had strabismus, whereas 3 of them had nystagmus. However, almost all of the patients showed a profound decrease in their vision because they were unable to pass the light and objects fixation test (Table 2). The mean spherical equivalent (SE) from the visual function examination by using the retinoscopy was -1.02 ± 2.47 and -0.88 ± 2.49 on the right and left eyes, respectively.

Table 2. Clinical Manifestations of Patients with CVI in the Department of Ophthalmology CMNH Kirana

Clinical Manifestations	Frequency (n = 16)
Strabismus	
ET 15°	4
XT 15°	1
XT 30°	3
Orthophoria	8
Nystagmus	
Yes	3
No	13
Visual Function	
LF (-) OF (-)	11
LF (+) OF (-)	3
LF (+) OF (+)	2

ET:esotropia, XT:exotropia, LF:light fixation, OF:object fixation

ERG, VEP, and CT scan or MRI was performed on 8 of 16 respondents. Based on the ERG results, six patients experienced a "severe retinal disorder," whereas, in the VEP examination, five of them experienced a decrease in amplitude and seven patients experienced a prolonged latency period (Table 3). Meanwhile, the results obtained from CT scans or MRI showed that cerebral atrophy was the most common disease.

Table 3. Electroretinography and Flash Visual Evoked Potential Test of Patients with CVI in the Department of Ophthalmology CMNH Kirana

Examination	Frequency (n=8)
Electroretinography test	
Mild	2
Moderate	0
Severe	6
Visual evoked potential test	
Amplitude	
Normal	3
Decreased	5
Latency	
Normal	1
Prolonged	7

Discussion

The median age of patients in this study was 20 months, but we observed a delay in growth and development in more than half of total patients. CVI can occur at various stages of development, namely in preterm/term infants, pre-school age children or school age, and adulthood. Cognitive, motoric, and speech deficits, as experienced by the patients with CVI, were very common abnormalities accompanied by central nervous system disorders.[9]

Ischemia and lack of oxygenation to the brain are the primary causes of CVI, which usually occur shortly after birth as the complications of preterm birth or difficulty in childbirth. A high incidence of CVI in developed countries is associated with their superior quality medical treatment that allows premature babies with severe brain damage to survive. However, in this study, all patients (n = 16) were term and as many as 12 patients were born by a normal birth. Brain hypoxia that occurs in most of these patients may not occur due to the complications of preterm birth, but rather due to a history of seizures. In this study, 14 patients had a history of previous seizures. Secondary complications due to seizures, metabolic diseases, hypoglycemia, and genetic syndromes can cause CVI. A study by Coady et al[10] reported that 53% of the patients with CVI were found to have a history of previous seizures.

The three most commonly observed comorbidities in this study were cerebral palsy, epilepsy, and intracranial infections. Suzanne et al[9] found several concomitant diseases, such as brain hypoxia due to cardiac/respiratory arrest, central nervous system infection, drug exposure, prematurity, cerebral palsy, and seizures/epilepsy, along with CVI. Coady et al[10] reported that cerebral palsy was the most common cause of CVI (26%).

Patients with CVI can show variations in the degree of visual acuity, visual disturbances, and fluctuations in visual performance. In cases of CVI with extensive damage, visual acuity and visual field can show severe degrees of disturbance.[9] As in this study, most patients showed a marked decrease in visual acuity, with as many as 11/16 patients were unable to fixate on light and objects. Other studies

in Tennessee obtained a similar result, where 40% of the patients had a perception of light but were unable to fixate.[11]

Retinoscopy was used to measure the refractive status, which displayed in the form of a spherical equivalent. Overall, there were more respondents with myopia. There are several considerations to be made when providing glasses to patients with brain damage. Generally, if the potential for increased visual acuity with glasses is lower than the potential visual acuity due to brain damage, glasses should not be given. On the other hand, in patients with hypermetropia or myopia, correction glasses should be given to improve vision.[12]

Strabismus could be the initial sign of CVI, especially if it is a constant exotropia. One possible cause of constant exotropia in the first year of life is due to abnormalities in the visual cortex. Apart from exotropia, esotropia also appears in patients with CVI and is an early sign of hydrocephalus or shunt malfunction, both of which are important causes of CVI. In this study, both types of strabismus were balanced. Vijay et al[11] and Nirajan et al[13] obtained different results with far greater amounts of exotropia (40% and 64%, respectively).

Only three respondents experienced nystagmus. Motor nystagmus is the most common type of strabismus that was found in patients with CVI. This type of nystagmus usually arises after damage to cortical control. Lowery et al[14] reviewed medical records of seven patients who had CVI. Unfortunately, only two of those had the ERG, and both showed a normal reading. In contrast to the results obtained by Lowery et al[14], eight of our patients had their ERG, and all of them showed an impaired retinal function. Although patients with CVI should show normal results on ERG examination, the abnormal ERG images observed in this study can be because of the underlying disease in patients with CVI, which can also cause damage to other areas such as the brain, chiasma, optic nerve, or the retina.[7]

In this study, the amplitude of flash VEP and its latency decreased in about 5/8 and 7/8 of patients, respectively. Other studies have found that a decrease in VEP occurs not only in flash VEP but also in sweep VEP.[15] Abnormal VEP results, below the normal average age, were commonly observed.[12]

Imaging can be used to establish CVI as a diagnosis because it can determine the underlying disease in the brain. It also can be used to predict the prognosis of the neuronal development of the child.[16] In this study, an MRI/CT scan of the head was performed on eight patients, and cerebral atrophy was found to be the most common pathology. Among all investigations, MRI is the best examination modality for detecting brain abnormalities.[17]

Conclusions

Clinical characteristics such as decreased vision, nystagmus, strabismus, accompanied by a history of prematurity and abnormalities in the central nervous system can help to establish diagnosis of CVI.

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