Ophthalmologists’ awareness of cerebral visual impairment – preliminary study

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Introduction. Cerebral visual impairment is the leading cause of severe visual impairment and blindness in infants and children in high-income countries. The main cause of this condition is damage of higher visual pathways and brain centers which leads to problems in processing visual information. In the last 20 years, an increase in the prevalence of cerebral visual impairment has been observed, but this diagnosis as a cause of vision problems in Serbia is not common. Objectives. The aim of this study was to assess the awareness of ophthalmologists of cerebral visual impairment. Methods. The Questionnaire for the assessment of cerebral visual impairment (Maitreya et al., 2018) was translated into Serbian, and used for data collection. The sample consisted of 45 ophthalmologists employed on the territory of the Republic of Serbia. Results. Two thirds of ophthalmologists from the sample self-assessed their knowledge as low (or nonexistent). However, the results showed that they had a satisfying level of awareness about cerebral visual impairment, and the majority of the participants correctly stated the most common risk factor for cerebral visual impairment. The highest awareness was observed in ophthalmologists who worked in health centers, those with more work experience, and especially those who primarily worked with children. Conclusion. Further informing the ophthalmologists about the diagnostic method of choice, symptomatology and differential diagnosis of cerebral visual impairment is desirable. Raising the awareness of doctors about this condition would lead to more frequent diagnoses, especially in babies with neurological impairments that are currently most likely misdiagnosed or unrecognized when it comes to their vision problems.

Keywords: cerebral/cortical visual impairment, children, ophthalmologists’ awareness

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Introduction

Many eye conditions (cataract, glaucoma, retinal dystrophy) result in vision problems which can be manifested as reduced visual acuity, restricted visual field and/or reduced contrast sensitivity. Apart from that, damage of higher visual pathways and brain centers causes problems in processing visual information. This condition is commonly known as cerebral visual impairment – CVI (Dutton, 2003), and it cannot be explained by any problem with eyes, or connecting nerve pathways.

Since the problem is located in the cortex, it was originally named cortical visual impairment. Over time, a broader term, cerebral visual impairment, emerged. The fact which is in favor of this term is that in children with this type of visual impairment, the condition is often accompanied by periventricular leukomalacia. This type of brain injury is located in white matter and, as a consequence, lesion of optic radiation appears commonly. Due to the fact that white matter and optical radiation are not part of the cortex, the term has changed from cortical to cerebral visual impairment. Concurrently, this term is more commonly used in Europe, while the term cortical vision impairment is frequently used by authors from the United States (Vučinić et al., 2019). All of the above has affected us to use the term cerebral visual impairment (CVI) in this paper. Regardless of terminological differences, the name cortical/cerebral visual impairment is used to refer to a condition leading to misinterpretation of the visual world (Dutton, 2003).

Cerebral visual impairment – causes and symptomatology

In the past two decades, study results indicate that CVI is the leading cause of severe visual impairment and blindness in infants and children in high-income countries (Gilbert et al., 2017; Good et al., 2001; Kozeis, 2010; Philip & Dutton, 2014), especially in prematurely born babies and children with cerebral palsy (Vučinić et al., 2019). It is also an emerging cause in low-income countries (Gilbert et al., 2017). Increasing prevalence of CVI in recent years could primarily be explained by better medical care, specifically perinatal care, and higher survival rates of children with serious neurological disorders (Kozeis, 2010; Matsuba & Jan, 2006; Philip & Dutton, 2014).

Cerebral visual impairment (CVI) is a neurological visual impairment caused by damage of gray and/or white matter of the brain (McKillop & Dutton, 2008). It refers to bilateral vision loss with a normal pupillary reaction and no abnormalities on eye examination (Dutton & Jacobson, 2001). CVI is an umbrella term that includes a broad range of visual deficits, from difficulties in visual perception to problems with visuomotor control (Goodale, 2013). Visual acuity of children with CVI can fluctuate during a day from blindness to emmetropia, and they can have a variety of associated visual field deficits.
Contrast sensitivity can be significantly reduced, while color vision is often intact. The main problem of children with neurological impairment in posterior visual pathways and/or the brain’s visual processing centers is receiving, integrating, interpreting and using visual information. Their brain has an inability to understand upcoming visual signals, and therefore, visual processing is delayed and inadequate (McKillop et al., 2006). This leads to a mismatch of incoming visual information.

It is important to highlight that present difficulties at the visual functions level, such as decreased visual acuity or visual field loss, can be explained by impairment of anterior (eye and optic nerve) or posterior visual pathways. Because of that, it is of greatest importance that ophthalmologists, neurologists or pediatricians have specific knowledge which can lead to a differential diagnosis between CVI and e.g. late visual maturation, high refractive errors, unusual retinal development, optic nerve abnormalities or oculomotor apraxia. The most common indicators of CVI are difficulties in recognizing objects and faces, problems with perception of moving objects in three-dimensional space, problems with orientation and mobility, strain with movement planning, difficulties handling complex visual scenes etc. (Dutton, 2003; Jablan & Stanimirov, 2011; McKillop et al., 2006). With this in mind, it is very important to notice any unusual visual behavior (Swaminathan & Patial, 2019).

Given the increase in the prevalence of CVI in developed countries, and since, on the other hand, this diagnosis as a cause of vision problems in Serbia is not common, the aim of this study was to assess the awareness of ophthalmologists of CVI in children.

**Methods**

**Participants**

**Table 1**

*Sample characteristics*

<table>
<thead>
<tr>
<th></th>
<th>n</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gender</td>
<td></td>
<td></td>
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<tr>
<td>male</td>
<td>14</td>
<td>31.0</td>
</tr>
<tr>
<td>female</td>
<td>31</td>
<td>69.0</td>
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<tr>
<td>Health care institution</td>
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<tr>
<td>primary</td>
<td>15</td>
<td>33.0</td>
</tr>
<tr>
<td>secondary</td>
<td>11</td>
<td>24.0</td>
</tr>
<tr>
<td>tertiary</td>
<td>19</td>
<td>42.0</td>
</tr>
<tr>
<td>Work experience</td>
<td></td>
<td></td>
</tr>
<tr>
<td>≤ 10</td>
<td>27</td>
<td>60.0</td>
</tr>
<tr>
<td>11–21</td>
<td>12</td>
<td>27.0</td>
</tr>
<tr>
<td>&gt; 21</td>
<td>6</td>
<td>13.0</td>
</tr>
<tr>
<td>Population</td>
<td></td>
<td></td>
</tr>
<tr>
<td>children</td>
<td>9</td>
<td>20.0</td>
</tr>
<tr>
<td>adults</td>
<td>36</td>
<td>80.0</td>
</tr>
</tbody>
</table>
The sample in this research was formed by the method of chain referral. The study involved 45 ophthalmologists employed on the territory of the Republic of Serbia, in primary, secondary and tertiary health institutions in Belgrade, Novi Sad, Kragujevac and Niš. A detailed description of sample characteristics is shown in Table 1.

As it can be seen from the Table 1, the majority of our sample consisted of women (almost 70%), employees in clinical centers (over 40%), ophthalmologists who primarily worked with adults (80%) and had less than 10 years of work experience (60%).

**Instrument and Procedure**

The *Questionnaire for the assessment of CVI awareness* (Maitreya et al., 2018) was translated into Serbian, and used for data collection. Before completing the questionnaire, ophthalmologists had to rate their knowledge about CVI.

The questionnaire consists of nine questions related to the etiology, symptoms, assessment, differential diagnosis, therapy and prognosis of CVI. Four answers are offered for each question, and participants should choose the answer which they consider correct. In line with the evaluation key, only correct answers were scored with 1 point. The original questionnaire has one item which directly indicates the number of diagnosed cases of CVI within one month. Considering the size of our country and the fact that this topic is still new, the aforementioned question was changed into: “*How many cases of CVI have you encountered during your practice?*” The reliability level of the questionnaire used was moderate (α = .69), but can be considered acceptable (Pallant, 2001, as cited in Daud et al., 2018).

The research was conducted online, through Google forms. Data was collected from December 2019 to March 2020. At the beginning of the questionnaire, an explanation was given about the purpose of the research and the content of the questionnaire. The participants gave their consent to participate in the study and then filled out the questionnaire.

This study was approved by the Ethical committee of the University of Belgrade – Faculty of Special Education and Rehabilitation (Decision No. 133/1).

**Statistical data analysis**

Descriptive measures and analysis of variance were used in statistical data analysis. Data was processed by Statistical Package for the Social Sciences (SPSS version 20.0). Before data processing, Levene’s test was used for assessing the equality of variances, and the significance of its score showed (in all cases) homogeneity of variances of subsamples (p > .05). Therefore, ANOVA was used to analyze the differences among means.
Results

First, the following question was analyzed: “How many cases of CVI have you encountered during your practice?” The results showed that more than half of the ophthalmologists from our sample had never diagnosed CVI ($n = 28$, 62.2%), 12 of them (26.7%) had seen less than five cases, one doctor (2.2%) chose the answer “from 5 to 10 cases diagnosed” and four participants said that they had diagnosed more than 10 patients with CVI.

This was followed by the rates of the ophthalmologists’ knowledge about CVI. The results are shown in Figure 1.

**Figure 1**

*Distribution of ophthalmologists’ self-assessed knowledge about CVI*

For the purpose of further analysis, these four groups were summarized into two categories – ophthalmologists who considered themselves to have knowledge about CVI (intermediate and high level, $n = 16$) and ophthalmologists who stated that they did not have knowledge about that diagnosis (very low and low, $n = 29$).

Correct statements on the questionnaire as a whole are presented in the Table 2.

The authors found that only one participant answered all questions from the questionnaire correctly, and that the minimum number of correct answers was two, as given by one participant. Five participants (11.1%) answered less than half of the questions correctly (three and four), while 38 participants (84.4%) gave more than half of the correct answers (five, six, seven and eight). Reviewing the number of correct statements on the questionnaire as a whole, the authors found that average number of correct answers was 6 ($M = 5.91$, $SD = 1.46$).
Subsequently, the analysis of particular answers was done. It was established that ophthalmologists considered that eye examination was necessary in this population, they knew what was meant by effective treatment for CVI and were well aware that management of CVI in children required multidisciplinary rehabilitative approach (n = 41, 91.1%). Furthermore, they expressed high knowledge about the most common risk factor for CVI and the cause of CVI in children (n = 38, 84.4%) as well. However, only half of the participants (n = 25, 55.6%) gave the correct answer to the following question: “What is the symptomatology of CVI in children?”. Additionally, less than half of the sample, i.e., 17 ophthalmologists (37.8%), answered correctly that MRI was the most adequate diagnostic method for CVI. The same number of participants believed that vision in children with CVI can (sometimes) be improved and also 17 of them (37.8%) answered correctly to the question regarding differential diagnosis of CVI in children. The smallest number of correct answers was obtained for the following question: “Which are the three most common causes of visual impairment among children in developed countries?” (n = 13, 28.9%).

After analyzing individual responses, the awareness level of the participants based on three monitored variables was examined: level of health care they worked in, work experience, and population they worked with.

Table 3 gives the mean scores of the correct answers of the participants employed in different health care institutions.

<table>
<thead>
<tr>
<th>Health care institution</th>
<th>n</th>
<th>M</th>
<th>SD</th>
<th>SEₘ</th>
</tr>
</thead>
<tbody>
<tr>
<td>Primary</td>
<td>15</td>
<td>6.40</td>
<td>1.06</td>
<td>.27</td>
</tr>
<tr>
<td>Secondary (hospitals)</td>
<td>11</td>
<td>5.82</td>
<td>1.17</td>
<td>.35</td>
</tr>
<tr>
<td>Tertiary (clinical centers)</td>
<td>19</td>
<td>5.58</td>
<td>1.81</td>
<td>.41</td>
</tr>
</tbody>
</table>

Table 2

Number of correct statements given by participants (N = 45)

<table>
<thead>
<tr>
<th>Number of correct answers</th>
<th>Number of participants</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>-</td>
</tr>
<tr>
<td>2</td>
<td>1</td>
</tr>
<tr>
<td>3</td>
<td>1</td>
</tr>
<tr>
<td>4</td>
<td>4</td>
</tr>
<tr>
<td>5</td>
<td>12</td>
</tr>
<tr>
<td>6</td>
<td>12</td>
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<tr>
<td>7</td>
<td>8</td>
</tr>
<tr>
<td>8</td>
<td>6</td>
</tr>
<tr>
<td>9</td>
<td>1</td>
</tr>
</tbody>
</table>
Based on the descriptive data shown in Table 3, it is clear that the participants employed in primary health care institutions had the highest level of awareness, while the awareness level of other two groups was similar. However, one way ANOVA showed that there were no statistically significant differences among ophthalmologists employed in different health care institutions ($F(2, 42) = 1.38, p = .26$).

The next step was to inspect whether there were statistically significant differences between ophthalmologists based on their work experience. The results are shown in Table 4.

**Table 4**

*Awareness of CVI based on work experience*

<table>
<thead>
<tr>
<th>Work experience</th>
<th>n</th>
<th>M</th>
<th>SD</th>
<th>SE_M</th>
</tr>
</thead>
<tbody>
<tr>
<td>≤ 10 years</td>
<td>27</td>
<td>5.81</td>
<td>1.57</td>
<td>.30</td>
</tr>
<tr>
<td>11 to 21 years</td>
<td>12</td>
<td>5.75</td>
<td>1.42</td>
<td>.41</td>
</tr>
<tr>
<td>&gt; 21 years</td>
<td>6</td>
<td>6.67</td>
<td>.82</td>
<td>.33</td>
</tr>
</tbody>
</table>

Result analysis showed that the participants with more than 21 years of work experience were those who had the highest awareness about CVI. The awareness level of other two groups was similar. Further analysis showed that ophthalmologists’ work experience did not significantly affect the awareness of CVI ($F(2, 42) = 0.93, p = .40$).

This was followed by the analysis of the ophthalmologists’ awareness of CVI based on the population they worked with. The results are shown in Table 5.

**Table 5**

*Awareness of CVI based on a population the ophthalmologists work with*

<table>
<thead>
<tr>
<th>Population</th>
<th>n</th>
<th>M</th>
<th>SD</th>
<th>SE_M</th>
</tr>
</thead>
<tbody>
<tr>
<td>Children</td>
<td>9</td>
<td>6.44</td>
<td>1.51</td>
<td>.50</td>
</tr>
<tr>
<td>Adults</td>
<td>36</td>
<td>5.78</td>
<td>1.44</td>
<td>.24</td>
</tr>
</tbody>
</table>

As shown in Table 5, the participants who primarily worked with children gave more correct answers on the questionnaire (which means higher awareness of CVI), compared to those who worked with adults. However, there were no statistically significant differences between the two groups ($F(1, 42) = 1.52, p = .22$).

Finally, the authors were interested in achievements on the questionnaire based on the self-assessed degree of knowledge about CVI. The results are shown in Table 6.
**Table 6**

*Awareness of CVI with regard to self-assessed knowledge about CVI*

<table>
<thead>
<tr>
<th>Self-rating</th>
<th>n</th>
<th>M</th>
<th>SD</th>
<th>SEM</th>
</tr>
</thead>
<tbody>
<tr>
<td>No knowledge</td>
<td>29</td>
<td>5.45</td>
<td>1.38</td>
<td>.26</td>
</tr>
<tr>
<td>Have knowledge</td>
<td>16</td>
<td>6.75</td>
<td>1.24</td>
<td>.31</td>
</tr>
</tbody>
</table>

Ophthalmologists who believed that they had knowledge of CVI had better results on our questionnaire, and ANOVA showed that there were statistically significant differences among those two groups of participants ($F(1, 42) = 9.86, p = .003$).

**Discussion**

Despite relatively low prevalence of childhood blindness (Kong et al., 2012), about 1.4 million of children all over the world are blind, while 19 million are visually impaired (Solebo & Rahi, 2014). Prevalence and causes of childhood visual impairment vary between countries, socioeconomic status, and health care (Resnikoff et al., 2016; Shah et al., 2011). The most frequent causes of childhood visual impairment/blindness in the world are retinal disorders, glaucoma, corneal opacity, cataract (Solebo & Rahi, 2014). In countries with higher income, the leading causes are CVI, retinopathy of prematurity and optic nerve hypoplasia (Solebo et al., 2017; Steinkuller et al., 1999).

It is necessary to raise awareness about this topic in Serbia, and this preliminary research is the first step towards that. Since only a third of the questioned ophthalmologists were aware of the fact that in developed countries CVI was the leading visual impairment in children, raising awareness of health professionals is important. The only effective treatment for children with CVI is multidisciplinary rehabilitative approach, which was recognized by majority of the participants.

Although two thirds of ophthalmologists from our sample self-assessed their knowledge of CVI as low (or non-existent), the results showed that they had a satisfying level of awareness about CVI (two thirds of them gave almost 70% of correct answers to the applied questionnaire). Also, majority of the participants stated perinatal hypoxia as the most common risk factor for CVI. Hypoxia/asphyxia on birth, with following neonatal encephalopathy is the main cause of mortality in newborns. In surviving newborns, it causes neurological disorders or neurodevelopmental impairments (Selimović et al., 2016; Vučinić et al., 2014). Frequency of perinatal hypoxia/asphyxia varies due to different definitions, but it can be said that in general the incidence ranges from 4-6%, depending on the course of pregnancy and time of childbirth (Jovandarić, 2018). Perhaps all children with neonatal encephalopathy as consequence of perinatal hypoxia should be considered at risk for developing CVI and, consequently,
should be monitored. This approach could be relevant, especially given that CVI is the cause of severe visual impairment in two out of 1000 term newborns and in as many as 19 out of 1000 children born between 20 and the 27th week of pregnancy (Vučinić et al., 2014). According to that fact, it can be assumed that there are many children with this condition in Serbia, but that they are most likely misdiagnosed or unrecognized.

The main cause of CVI is damage of visual cortex (five different areas from V1 to V5, based on function and structure) which are involved in the integration and interpretation of visual information (Goodale, 2013; Huff et al., 2019), or damage of the white matter of periventricular area, accompanied with severe form of focal necrosis (Folkerth, 2006). Despite the correct answers about the main cause of CVI, 50% of ophthalmologists from the sample believed that visual evoked potentials (VEP) was the diagnostic method of choice. Since methods based on evoked potentials (EP) enable the registration of electrical potentials of nervous tissue, VEP enables the detection of nerve pathway damage, i.e., damage to the optic nerve. Therefore, in order to confirm damage of cortical structures, the method of choice should be MRI. Certainly, the question arises why the participants did not opt for the appropriate diagnostic method, after they gave the correct answer on causes of CVI?

Clinical symptoms of CVI were familiar to 55% of the examined ophthalmologists, and 45% of the participants were not familiar with difficulties indicating CVI. Although they are the first “stop” when it comes to diagnosis and treatment of vision problems, it is necessary for these specialists to be experienced in order to suspect this condition, i.e., to avoid overlooking it. Differential diagnosis between amblyopia, delayed visual development, autism and CVI requires good knowledge of clinical symptoms. In that context, it is important to emphasize that CVI is always bilateral, while amblyopia is usually unilateral. As for delayed development of visual abilities, children with this problem develop normal visual functions by the end of the first year, while this does not happen in children with CVI. Unusual visual behavior can be seen in children with autism, but the diagnostic criteria is visual acuity the values of which are average in children with autism, while in children with CVI there is a noticeable fluctuation in visual abilities (Maitryea et al., 2018). Apart from the above symptomatology, only MRI can remove any doubt.

After analyzing the relationship between the monitored variables, significant differences in the level of ophthalmologists’ awareness based on health care institutions, work experience and the population they worked with were not found. The highest awareness was observed in ophthalmologists who worked in health centers, those with more work experience, and especially those who primarily worked with children. The authors believe that the obtained information should be commented. The first eye examination is usually performed in primary eye care institutions, and the significance of this finding
is clear. Another interesting fact is that work experience related to knowledge/awareness of CVI confirms the importance of continuous education and practical experience. Finally, the fact that pediatric ophthalmologists have higher level of awareness of CVI is a clear confirmation that this diagnosis is becoming more common in children, while in adults it is still insufficiently researched. Given the observed increase of survival rate of children with neurological disorders in the last 20 years, it is a probable assumption that there are a number of young adults with significant visual problems which are not recognized and monitored, most likely due to other neurological, cognitive, behavioral or communication difficulties they have.

Finally, it should be emphasized that ophthalmologists had very high and objective self-assessment of CVI awareness. It was confirmed that those who believed to have a high level of knowledge, also had higher awareness about this topic (a significantly better score on the Questionnaire), compared with their colleagues who had the opposite opinion. The findings definitely indicate the existing awareness of doctors about this neurological condition which can affect the quality of life and everyday participation of children and their families. However, the sample size does not allow generalization of the results. Nevertheless, according to the results of this pilot study, further informing the ophthalmologists about the clinical symptomatology and differential diagnosis of CVI is desirable.

Conclusion

The development of prenatal and perinatal care has led to the increased number of preterm infants surviving, but premature birth often leaves consequences. Neurological impairment and damage to the brain visual areas which is a result of CVI are increasingly present, especially in developed countries. Although this condition is becoming more common, a significant number of children with the aforementioned diagnosis have not been noticed in Serbia. In order to recognize and include these children in adequate treatment, it is necessary for neonatologists, pediatricians, neurologists, physiatrists, special educators and other specialists who work with children with neurological issues, to be familiar with symptomatology, differential diagnosis and diagnostic methods. Since unusual visual behavior is the most common symptom of CVI, ophthalmologists should primarily be informed about CVI guidelines, because early detection is crucial for effective rehabilitative treatment.
References


Informisanost oftalmologa o cerebralnom oštećenju vida – preliminarno istraživanje

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Ključne reči: cerebralno/kortikalno oštećenje vida, deca, informisanost oftalmologa

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