Eye (2021) 35:1191–1197
https://doi.org/10.1038/s41433-020-1101-z

The ROYAL COLLEGE of OPHTHALMOLOGISTS

ARTICLE

# Causes of severe visual impairment in infants and methods of management

Zuhal Ozen Tunay[1] · Zeynep Ustunyurt[2] · Aysun Idil[3]

Received: 18 May 2020 / Revised: 7 July 2020 / Accepted: 9 July 2020 / Published online: 16 July 2020


## Abstract

**Objectives** To examine the causes of severe visual impairment (SVI) in infants below the age of 2 years and to review management methods.

**Methods** The data of 2657 patients followed between January 2014 and July 2019 were reviewed, and 148 (5.6%) infants who had SVI were enrolled. Data including age, gender, affected anatomical site, diagnosis, presence of any non-ophthalmological deficiencies, and methods of management were reviewed. The diagnoses were investigated in the categories of avoidable and unavoidable basis. The methods of management were analysed from the perspective of low vision habilitation.

**Results** The mean age at first eye examination was 6.61 ± 5.25 months, and 84 (56.7%) infants were male. Of the 148 infants, 69 (46.6%) were premature. Cerebral visual impairment (CVI) was the most common diagnosis in both preterm (39.1%) and term (11.4%) infants. Delayed visual maturation, optic nerve pathologies, oculocutaneous albinism, and congenital cataract were the other frequent causes. The rate of multiple disabilities was 30% in the whole group and 94% in infants with CVI. Most of the babies had a normal-appearing globe (43.3%). Retina was affected in 23.7% of the infants. Avoidable causes were identified in 79.7% of the infants. The used methods of management were optic interventions, visual stimulation therapy, medical and/or surgical treatment.

**Conclusions** CVI was found the most common cause of SVI in both preterm and term-born infants, and the higher rate of multiple disabilities in these infants was remarkable. Optic interventions and visual stimulation therapy were the most common methods of management.

## Introduction

Severe visual impairment (SVI) in infants and children can negatively affect the educational and psychosocial life of the individuals, and it impairs the quality of life. Besides this, it has social, economic and psychological impacts on their families and the community [1, 2].

The researches in infants (0–2 years of age) with SVI are limited compared to the other age groups due to the challenges in the evaluation of vision and diagnosis of the impairment, and the presence of comorbidities [3]. The prevalence, causes, and the way of presentation vary between different parts of the world. Early diagnosis and treatment of some diseases such as Vitamin A deficiency, congenital cataract, retinopathy of prematurity (ROP), and retinoblastoma, can prevent blindness and even save the life of the patient [4–6]. Early diagnosis and treatment (if possible) are also important to minimize the avoidable causes of SVI [2, 4]. For this purpose, in this study, we analysed the causes of the SVI in the individuals 0–2 years of infantile age and we reviewed the methods of management.

## Subjects and methods

Ethical approval for this study was obtained from the Clinical Trials Ethics Committee (Ministry of Health, Zekai

✉ Zuhal Ozen Tunay
zuhaltunay@gmail.com

[1] University of Health Sciences, Zekai Tahir Burak Education and Research Hospital, Department of Ophthalmology, Ankara, Turkey

[2] University of Health Sciences, Zekai Tahir Burak Education and Research Hospital, Department of Developmental Pediatrics, Ankara, Turkey

[3] Ankara University Faculty of Medicine, Department of Ophthalmology, Low Vision Rehabilitation and Research Center, Ankara, Turkey

Tahir Burak Education and Research Hospital Ethics Committee, Registration number: 23.07.2019-110/2019). All works were conducted in accordance with the Declaration of Helsinki and with the approval of Institutional Review Board.

Our tertiary level hospital is located in Ankara, the capital city of Turkey, and the city has a population of over 5.5 million people. Our hospital has the biggest neonatal intensive care unit of Turkey with a capacity of 150 incubators and serves thousands of babies from Ankara and the surrounding Anatolian region. Since very few centres in our country provide low vision support for infants, our study population involved infants who referred to our Ophthalmology Clinic by paediatricians and ophthalmologists from all Anatolian regions.

We retrospectively reviewed the data of 2657 infants (<2 years of age) followed in our clinic between January 2014 and July 2019, and we included the infants with SVI in the study. The infants with a unilateral visual impairment were excluded. All infants underwent a full ophthalmologic evaluation including visual and orthoptic assessment, anterior and posterior segment examination, and cycloplegic refraction, by the same ophthalmologist (Dr. Tunay). Visual acuity was examined with the Lea Gratings Preferential Looking Test for the assessment of detection grating acuity. The Standard test distance was 57 cm (~2 feet) from the infant's face. At this distance, one centimetre equals one degree of visual angle, and the number of cycles/cm (cpcm) corresponds to grating acuity as cycles per degree (cpd). For infants who do not respond to grating test card placed at 57 cm distance, the gratings were held at 30 cm and/or 15 cm distance (the half or quarter of the original distance), and the grating acuity values were calculated according to distance formula [(distance used/57 cm) X cpcm = cpd]. The fixation pattern and visual evoked potentials (VEP) were evaluated for the infants whose visual acuity could not be measured with Lea gratings. Signs of low vision such as roving eye movements, oculodigital reflex, or nystagmus were also recorded. The infants who had no light perception, the infants who over 2-month-old and who had only light perception, the infants who had no central fixation and who had signs of very poor vision (like searching nystagmus, ocular bobbing, or oculodigital massage), the infants over 6-month-old and who had a fixation less than five- seconds and no pursuit were diagnosed with SVI. Preterm babies were evaluated according to the gestational-week-adjusted corrected age.

The prism cover test or Hirschberg and Krimsky tests were performed for orthoptic examination, depending on the cooperation status of the baby. A baseline refraction measurement was done with binocular autorefractor (Plusoptix A09; Plusoptix GmbH, Nuremberg, Germany). A hand-held portable slit lamp biomicroscope (Shin-Nippon, Rexxam Co, Tokyo, Japan) was used for anterior segment examination. Cycloplegic retinoscopy was performed for the detection of refractive errors. The posterior segment was examined with a binocular indirect ophthalmoscope and with ultrasonography when needed. The infants who had SVI without any ocular pathology were evaluated for other causes. Neurological examination, VEP, and cranial imaging with ultrasonography and/or magnetic resonance imaging were performed when required. The diagnosis of "cerebral visual impairment (CVI)" was based on 3 criteria according to these clinical and imaging findings: reduced visual acuity, a normal ocular examination, and confirmation of damage to the posterior visual pathways. The visually impaired infants who had a normal ophthalmological, cerebral and neurological examination, were diagnosed as "delayed visual maturation" if they showed age-appropriate improvement of visual acuity in subsequent examinations.

Data including age at the first appointment (month), gender, family history, affected anatomical site, cause of SVI (the primary cause of SVI in the less affected eye was selected in the presence of multiple diagnoses) and chosen methods of management was evaluated. The presence of any non-ophthalmological deficiencies (e.g. psychomotor, mental or hearing) was recorded. Classification of the affected anatomic site was set up as, cornea, lens, uvea, retina, optic nerve, whole globe and a normal-appearing globe. The causes of SVI were also classified as avoidable and unavoidable causes. Treatable and preventable/partially preventable causes were analysed under the classification of avoidable causes. Treatable causes include the diagnoses that we have a chance to treat the condition with optical, medical and/or surgical methods. Preventable/partially preventable causes involve conditions (e.g., intrauterine infections, traumatic birth and hereditary diseases which may be related to consanguineous marriages) that can be reduced by the improvement of primary health care services. Cerebral visual impairment and ROP were also classified as potentially preventable through perinatal and neonatal health care precautions.

The methods used for management and low vision habilitation were evaluated as the following categories: optic interventions, medical treatment, surgical treatment, and low vision habilitation with visual stimulation therapy. Eyeglasses, contact lenses and photochromic filters were classified in the category of optic interventions. Systematic and sequential uses of visual stimuli such as lights, bright colours, and high contrast black-and-white patterns were performed for visual stimulation therapy.

Statistical analyses were performed using SPSS version 16.0 (SPSS Inc., Chicago, IL). Frequency (percent) for categorical variables, mean±standard deviation and median value for metric variables was given as descriptive statistics.

## Results

Totally 157 infants between 0 and 24 months of age were diagnosed with SVI in the study period. Nine of them had unilateral SVI and excluded from the study group (3 unilateral congenital cataracts, 2 microphthalmos, 1 anophthalmos, and unilateral phthises secondary to ROP, familial exudative vitreoretinopathy or persistent foetal vasculature). As a result, 148 infants were included in the study. The mean age at first eye examination was 6.61 months with a standard deviation (SD) of 5.25 months, and the median age was 6 months. Sixty-nine babies (46.6%) were born preterm before 37 weeks of gestational age. The gender ratio (male:female) was 1.31, and 84 (56.7%) of the infants were male. Most of the patients (93 infants, 62.8%) had come from several regions for visual assessment, treatment or habilitation.

Visual acuity was evaluated with Lea grating acuity cards in 107 (72.3%) infants. The median value and the range in cpd (decimal) was 1.0 cpd (0.032) and 0.125–2.4 cpd (0.004–0.08), respectively. Visual acuity could not be measured in 41 (27.7%) infants who had no central fixation and who had signs of very poor vision, like searching nystagmus, ocular bobbing or oculodigital massage. The vision was assessed with light perception and VEP for these infants. Only light perception response was obtained in 26 (17.6%) infants who had no central fixation, whereas light perception could not be detected in 15 (10.1%) infants. VEP amplitudes could not be recorded in 19 (12.8%) infants.

The rate of parental consanguineous marriages was 18.2%. Positive family history for the same disease was present in 21.6% of the study population. The rate of associated non-ophthalmological deficiencies (e.g. motor, mental and hearing) was 30.4%, and this rate was highest in the patients with CVI (34 of 36 infants, 94.4%).

The distribution of diagnoses was shown in Table 1. The most frequent diagnosis was CVI (36 infants, 24.3%), and 27 of the 36 infants with CVI were prematurely born. The causes of CVI were periventricular leukomalacia, hydrocephalus, hypoxic encephalopathy and Grade 4 intracranial haemorrhage (38.9%, 33.3%, 16.7% and 11.1%, respectively).

**Table 1** Distribution of main pathologies leading to severe visual impairment.

| The pathology leading to severe visual impairment | Number of term-born infants | Number of preterm infants | Total number of infants | Total infants (%) |
|---|---|---|---|---|
| Cerebral visual impairment | 9 | 27 | 36 | 24.3 |
| Delayed visual maturation | 5 | 10 | 15 | 10.1 |
| Refractive errors | 6 | 7 | 13 | 8.8 |
| Congenital cataract | 8 | 3 | 11 | 7.4 |
| Oculocutaneous albinism | 8 | 3 | 11 | 7.4 |
| Optic atrophy | 4 | 5 | 9 | 6.2 |
| Retinopathy of prematurity (Stage 5) | 0 | 5 | 5 | 3.4 |
| Familial exudative vitreoretinopathy | 5 | 0 | 5 | 3.4 |
| [a]Anatomic pathology of eye and globe | 4 | 1 | 5 | 3.4 |
| Optic disc hypoplasia | 1 | 3 | 4 | 2.7 |
| Cone distrophy | 4 | 0 | 4 | 2.7 |
| Congenital glaucoma | 3 | 1 | 4 | 2.7 |
| Idiopathic nystagmus | 3 | 1 | 4 | 2.7 |
| Metabolic diseases | 2 | 2 | 4 | 2.7 |
| Choroidal coloboma | 3 | 0 | 3 | 2.0 |
| Corneal opacity/scar | 2 | 1 | 3 | 2.0 |
| Shaken baby syndrome | 2 | 1 | 3 | 2.0 |
| Retinoblastoma | 3 | 0 | 3 | 2.0 |
| Leber congenital amaurosis | 2 | 0 | 2 | 1.35 |
| Optic disc coloboma | 2 | 0 | 2 | 1.35 |
| Coats disease | 1 | 0 | 1 | 0.7 |
| Foveal aplasia | 1 | 0 | 1 | 0.7 |
| Total | 79 | 69 | 148 | 100.0 |

[a]Anatomic pathology of eye and globe: anophthalmos, microphthalmos, nanophthalmos.

We analysed the distribution of primary causes of SVI in preterm and term-born infants separately. Cerebral visual impairment was the most common diagnosis in both preterm (39.1%) and term (11.4%) infants. Delayed visual maturation was the second most common diagnosis in the whole group, and all of the babies in this group showed age-appropriate development of vision with their peers during the follow-up period. Oculocutaneous albinism and congenital cataract were the other common causes in term-born infants. However, refractive errors and optic nerve pathologies were the other frequent causes in preterm infants (Table 1).

High refractive errors [hyperopia >6 dioptres (D), myopia >6 D, astigmatism >3 D] were the main cause of SVI in 13 (8.8%) babies. Of them, 6 infants had hyperopia, 5 had myopia and 2 had astigmatism [mean ± SD (minimum, maximum): +8.25 ± 1.08 (+7.00, +10.00) D; −9.60 ± 1.63 (−7.50, −12.00) D; 4.75 ± 0.35 (4.50, 5.00) D, respectively]. Two of the patients with high refractive errors had Duane syndrome, one had Goldenhar syndrome and one had Cornelia de Lange syndrome. Accompanying refractive errors were present in 71 (48%) of the infants who had different main diagnoses. The spherical equivalent was +1.71 D, the median value was 2.00 D, and the range was between −6.00 D and +4.50 D.

Of the 9 babies with optic atrophy, 4 had craniosynostosis and 4 had the history of intracranial pathologies. De Morsier syndrome was diagnosed in an infant with optic disc hypoplasia.

The distribution of affected anatomic site was shown in Table 2. Most of the babies had normal-appearing globe (43.3%). Retina (23.7%) was the most affected anatomical region, and it was followed by optic nerve (10.1%) and lens pathologies (7.4%).

Treatable and preventable/partially preventable causes were classified as avoidable reasons and this classification was shown in Table 3. Avoidable causes were identified in 79.7% of the infants.

The methods of management for visually impaired babies in this study group were listed in Table 4. More than one method was applied in most of the babies. Visual stimulation therapy was performed for 84.5% of the study population. Optic interventions were required for 69.6% of the infants and the eyeglasses for refractive errors were the most popular one. Photochromic filters were used for the patients with photophobia, hereditary retinal pathologies and low contrast sensitivity. Medical treatment was applied for the patients with corneal disease, metabolic diseases and congenital glaucoma (before and sometimes after the surgical therapy if required). Surgery was performed for the patients with following diagnoses: congenital cataract, congenital glaucoma, ROP, familial exudative vitreoretinopathy, corneal pathology, persistent foetal vasculature, retinoblastoma and coats disease.

**Table 2** Anatomic site of abnormality leading to severe visual impairment.

| Anatomic site | *n* | % |
|---|---|---|
| [a]Normal-appearing globe | 64 | 43.3 |
| Retina | 35 | 23.7 |
| Optic nerve | 15 | 10.1 |
| Lens | 11 | 7.4 |
| [b]Whole globe | 9 | 6.1 |
| Uvea | 3 | 2.0 |
| Cornea | 3 | 2.0 |
| [c]Other | 8 | 5.4 |
| Total | 148 | 100.0 |

[a]Cerebral visual impairment, delayed visual maturation, refractive errors.

[b]Anophthalmos, microphthalmos, nanophthalmos, congenital glaucoma.

[c]Metabolic diseases, idiopathic nystagmus.

**Table 3** Avoidable causes of severe visual impairment.

| Causes | *n* | % |
|---|---|---|
| Avoidable causes | 118 | 79.7 |
| Treatable | 35 | 23.6 |
| Refractive errors and amblyopia | 13 | 8.8 |
| Congenital cataract | 11 | 7.4 |
| Congenital glaucoma | 4 | 2.7 |
| Corneal opacity/scar | 3 | 2.0 |
| Retinoblastoma | 3 | 2.0 |
| Coats disease | 1 | 0.7 |
| Preventable/partially preventable | 83 | 56.1 |
| [a]Cerebral visual impairment | 36 | 24.3 |
| Delayed visual maturation | 15 | 10.1 |
| Genetic and hereditary causes | 11 | 7.4 |
| Secondary optic atrophy | 9 | 6.2 |
| [a]Retinopathy of prematurity | 5 | 3.4 |
| Metabolic diseases | 4 | 2.7 |
| Shaken baby syndrome | 3 | 2.0 |
| Unavoidable causes | 30 | 20.3 |
| Oculocutaneous Albinism | 11 | 7.4 |
| [b]Anatomic pathology of globe | 5 | 3.4 |
| Colobomas | 5 | 3.4 |
| Idiopathic infantile nystagmus | 4 | 2.7 |
| Optic disc hypoplasia | 4 | 2.7 |
| Foveal aplasia | 1 | 0.7 |
| Total | 148 | 100.0 |

[a]Partially preventable with improvement of neonatal and perinatal care. Early diagnosis and management is also important.

[b]Anatomic pathology of globe: anophthalmos, microphthalmos, nanophthalmos.

**Table 4** Methods of management used for infants with severe visual impairment (more than one method was applied for most of the infants).

| Method of management | *n* | % |
|---|---|---|
| Optic interventions | 103 | 69.5 |
| Eyeglasses | 84 | 56.7 |
| Photochromic filter glasses | 24 | 16.2 |
| Contact lenses | 6 | 4.1 |
| Medical treatment | 11 | 7.4 |
| Surgical treatment | 29 | 19.6 |
| Visual stimulation therapy | 125 | 84.5 |

## Discussion

In the literature, there are many reports which examine the causes of SVI in childhood, but very few numbers of them include infants under the age of 2 years [2, 5, 6]. Difficulties in the evaluation of vision and the presence of comorbidities are the main compelling factors in this age group [3]. Nevertheless, early diagnosis and management are so important, especially for treatable causes such as congenital cataract, congenital glaucoma, or ROP [5, 6]. In addition, functional vision may be improved with visual stimulation therapy in the first months of life in babies with CVI and delayed visual maturation [7–9]. For all these reasons, we thought it significant to examine the causes of SVI in infants to develop effective strategies for early diagnosis and management. So, we analysed the causes of SVI and the methods of management in a cohort of infants aged 0–24 months in this study.

The distribution of diagnosis in individuals with low vision and blindness vary between the countries [4, 5, 10]. In the developed world, the major causes of SVI are ROP, CVI and optic nerve diseases [5, 11–14]. In recent years, the primary causes in some developing regions have been shifting from cataracts, infection, malnutrition and uncorrected refractive errors to more similar causes seen in developed countries [6, 15–17]. The most common diagnosis in our study was CVI (24.3%), as in studies from many countries. The possible reasons for this high rate of CVI may be increased survival of preterm infants, advances in neuroimaging and increased awareness of this problem [5, 11–13]. Bosch et al. and Solebo et al. also emphasized the increasing prevalence of CVI and the importance of perinatal health [5, 18]. Blindness due to an event in the perinatal period was seen 30% of the patients in studies from Niger and Turkey [2, 19]. Chhablani and Kekunnaya declared that preterm birth might lead to many neuro-ophthalmological problems and especially CVI [20]. The rate of cerebral-origin visual problems may be decreased with the improvement of perinatal health care for mothers and babies [2, 18, 20].

In our study, more than 30% of visually impaired infants had additional non-ophthalmological deficiencies and this rate was more than 90% in the patients with CVI. This finding corresponds well with the studies from Sweden and Norway. In these studies, the prevalence of additional disabilities was found over 50%, and it was more than 80% in children with neuro-ophthalmological diseases such as CVI [21–23]. These findings emphasize the importance of multidisciplinary evaluation of these children [9, 12, 21–23].

Retinopathy of prematurity was not a common cause of SVI, despite the great proportion of premature infants in our study population. The reduction in the prevalence of ROP-related blindness in the last decade was mentioned in another study from Turkey [2]. The improvement of neonatal care support and screening programmes for ROP in Turkey were effective in controlling ROP-related visual impairment [2, 9]. Globally, ROP is the primary threat to vision in preterm babies [10, 24, 25]. However, CVI became a more frequent cause of SVI for a preterm infant in high-income countries, in recent years [9, 11, 13–15].

In this study, main cause of SVI was high refractive errors in almost 10% of infants. On the other hand, nearly 50% of the study population with any diagnosis had required glasses because of refractive errors. The rate of refractive errors among the visually-impaired children varies between 7 and 42% according to the age-group of study population and development level of the region [4, 6, 15, 19, 26, 27]. Correction of refractive errors in the first years of life is important to overcome refractive amblyopia [7, 9, 10].

Zheng et al. investigated the changing profile of paediatric optic atrophy patients over the years and they reported the increased importance of prematurity-related complications and perinatal events. They pointed out the medical comorbidities in patients with optic atrophy. The most frequents comorbidities were hydrocephalus (20%), developmental delay (17%), seizures (16%) and cerebral palsy (14%) in their study [28]. Optic atrophy was the primary cause of SVI in 5.1% of term-born infants and 7.2% of preterm infants in our study. The most common comorbidity was hydrocephalus in preterm infants and seizures in term-born peers.

Avoidable causes, including treatable and preventable/partially preventable ones, were determined nearly 80% of the infants in this study. We encountered the patients with CVI in the category of partially preventable causes since the frequency can be reduced by prenatal and perinatal care and precautions [2, 4, 5]. If the infants with CVI were thought in the group of unavoidable causes, then the frequency of avoidable causes decrease to the level of 55%. In previous studies, the frequency of avoidable causes varies between 18 and 78% according to study population, socioeconomic status of society and the method of classification [5, 6, 10, 11]. In a study from western part of our country, the cerebral causes were not taken into account in a group of avoidable causes and the frequency of avoidable causes was found 62% similar

to our study [2]. In some previous studies, the rate of avoidable causes was found 77.8% in Indonesia, 69.9% in Eritrea and 69.4% in Congo [26, 27, 29].

The retina was the most common affected anatomic site in visually-impaired children in many previous studies. However, most of the visually-impaired children had a normally-appearing globe in recent studies from developed countries. In the studies from United Kingdom, Norway and Holland, in which the CVI was found the leading cause of childhood SVI, the most of the patients had normally-appearing globe (48, 37 and 29%) [11, 12, 21]. Retina was the most common involved site in the studies from Iran (62.6%), Brazil (42.1%), Malaysia (33.0%) and China (24.9%) [29–32]. On the other hand, lens and cornea were the most affected anatomical sites of involvement in the studies from low-income regions [6, 12]. In our study, 43% of the infants had a normal-appearing globe similar to the studies from European countries. The retina was the primary site of involvement in 23.7% of infants similar to the studies from the west region of Turkey (24%) and China (24.9%) [2, 17].

The male:female ratio in our study was 1.31, similar to many published studies with male predominance [2, 5, 22–26]. The rate of parental consanguineous marriages was 18.2%, and positive family history was 21.6%. These high rates are important for genetic-based diseases and genetic counselling [2, 6, 10].

We also mentioned our methods of management for visually-impaired infants in this study. We used optic interventions for more than two-thirds of the babies primarily for refractive errors. We wanted to emphasize the importance of determining the refractive errors correctly in infants with low vision. Even in the adult population with low vision, the correction of refraction with conventional eyeglasses without any other more sophisticated and expensive methods provide useful functional vision in ~20% of the patients [1, 9, 33]. Thus the refractive and accommodative problems should be checked carefully. We used photochromic filters in 16% of the babies, mostly for oculocutaneous albinism and hereditary retinal diseases. We used them for control of photophobia and enhancement of contrast sensitivity, as mentioned in the literature [1, 17]. We performed visual stimulation therapy for more than 80% of the infants in this study. Alimovic et al. showed that the visual stimulation programme helped the improvement of visual functions in babies [8]. However, one of the challenging points is the assessment of the effect of visual stimulation therapy. Many variables like physiologic improvement of vision with advancing age and changes in the course of the main disease may affect visual acuity. Therefore, to assess the visual acuity with before-and-after studies may not be a suitable approach for the infants [34].

The major limitations of this study are that it is clinically based and retrospective. However, all the infants had been examined by the same ophthalmologist. This condition may overcome the limitation of poor standardization because of multiple examiners. On the other hand, since low vision support for infants is provided by a very few centres in our country, infants in this study were referred to our Ophthalmology Clinic from all Anatolian regions. Because of this, although our findings are not representative of Turkey, we believe that they may contribute to the understanding of the causes of SVI in infants. Another limitation of this study is that the primary pathology with the major effect on vision was taken into account in cases with multiple ophthalmologic problems, since the common coexistence of more than one cause may affect the results. Nonetheless, this study includes a very specific and challenging age group (<2 years of age). In this context, we hope that it will contribute to the literature.

Finally, our study gives information on the causes of SVI in infants under the age of 2 years and our approach to managing. One of the striking points in this study is a great rate of CVI in both preterm and term-born infants, and the higher rate of multiple disabilities in these infants. The knowledge of the causes of SVI and changing trends over time may contribute to identifying efficient intervention strategies. Further researches for the advancement of these strategies may include the development of screening programmes for treatable causes, a multidisciplinary approach for infants with multiple disabilities, genetic counselling for hereditary and familial disorders, and improvement of perinatal care.

## Summary

### What was known before

- The knowledge of the causes of visual impairment in infants and children, and changing trends over time may contribute to identifying efficient intervention strategies for public health. The prevalence, causes, and the way of presentation vary between different parts of the world.

### What this study adds

- Cerebral visual impairment is the most common cause of severe visual impairment in both preterm and term-born infants (<2 years of age) in this study, and the higher rate of multiple disabilities in these infants is remarkable.

## Compliance with ethical standards

**Conflict of interest** The authors declare that they have no conflict of interest.

**Publisher's note** Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

## References


1. Ozen Tunay Z, Caliskan D, İdil A, Oztuna D. Clinical characteristics and low vision rehabilitation methods for partially sighted school-age children. Turk J Ophthalmol. 2016;46:68–72.
2. Ozturk T, Er D, Yaman A, Berk AT. Changing trends over the last decade in the aetiology of childhood blindness: a study from a tertiary referral centre. Br J Ophthalmol. 2016;100:166–71.
3. Matsuba CA. The challenges of research in children with VI. Dev Med Child Neurol. 2017;59:674.
4. Gilbert CE, Foster A. Childhood blindness in the context of VISION 2020: the right to sight. Bull World Health Organ. 2001;79:227–32.
5. Solebo AL, Teoh L, Rahi J. Epidemiology of blindness in children. Arch Dis Child. 2017;102:853–7.
6. de Paula CH, Vasconcelos GC, Nehemy MB, Granet D. Causes of VI in children seen at a university-based hospital low vision service in Brazil. J Aapos. 2015;19:252–6.
7. Hensch TK, Quinlan EM. Critical periods in amblyopia. Vis Neurosci. 2018;35:1–4.
8. Alimovic S, Katusic A, Mejaski-Bosnjak V. Visual stimulations' critical period in infants with perinatal brain damage. NeuroRehabilitation. 2013;33:251–5.
9. Özen Tunay Z. Low vision, sequela and rehabilitation in retinopathy of prematurity. Curr Retin. 2018;2:81–7.
10. Kong L, Fry M, Al-Samarraie M, Gilbert C, Steinkuller PG. An update on progress and the changing epidemiology of causes of childhood blindness worldwide. J Aapos. 2012;16:501–7.
11. Boonstra N, Limburg H, Tijmes N, van Genderen M, Schuil J, van Nispen R. Changes in causes of low vision between 1988 and 2009 in a Dutch population of children. Acta Ophthalmol. 2012;90:277–86.
12. Rahi JS, Cumberland PM, Peckham CS, British Childhood VI Interest Group. Improving detection of blindness in childhood: the British Childhood Vision Impairment study. Pediatrics. 2010;126:e895–903.
13. Pham C, Sheth SJ, Keeffe JE, Carden SM. New trends in childhood vision impairment in a developed country. J Aapos. 2017;21:496–8.
14. Jonas DE, Amick HR, Wallace IF, Feltner C, Schaaf EBV, Brown CL, et al. Vision screening in children aged 6 months to 5 years: evidence report and systematic review for the US Preventive Services Task Force. JAMA. 2017;318:845–58.
15. Flaxman SR, Bourne RRA, Resnikoff S, Ackland P, Braithvaite T, Cicinell MV, et al. Global causes of blindness and distance vision impairment 1990–2020: a systematic review and meta-analysis. Lancet Glob Health. 2017;5:1221–34.
16. Uprety S, Khanal S, Morjaria P, Puri LR. Profile of paediatric low vision population: a retrospective study from Nepal. Clin Exp Optom. 2016;99:61–5.
17. Gao G, Yu M, Dai J, Xue F, Wang X, Zou L, et al. Demographic and clinical characteristics of a paediatric low vision population in a low vision clinic in China. Clin Exp Optom. 2016;99:274–9.
18. Bosch DG, Boonstra FN, Willemsen MA, Cremers FP, de Vries BB. Low vision due to cerebral VI: differentiating between acquired and genetic causes. BMC Ophthalmol. 2014;14:59.
19. Aghaji A, Okoye O, Bowman R. Causes and emerging trends of childhood blindness: findings from schools for the blind in Southeast Nigeria. Br J Ophthalmol. 2015;99:727–31.
20. Chhablani PP, Kekunnaya R. Neuro-ophthalmic manifestations of prematurity. Indian J Ophthalmol. 2014;62:992–5.
21. Haugen OH, Bredrup C, Rødahl E. Visual impairment in children and adolescents in Norway. Nedsatt syn hos barn og unge i Norge. Tidsskr Nor Laegeforen. 2016;136:996–1000.
22. Blohmé J, Tornqvist K. Visual impairment in Swedish children. II. Etiological factors. Acta Ophthalmol Scand. 1997;75:199–205.
23. Blohmé J, Tornqvist K. Visual impairment in Swedish children. III. Diagnoses. Acta Ophthalmol Scand. 1997;75:681–7.
24. Blencowe H, Lawn JE, Vazquez T, Fielder A, Gilbert C. Preterm-associated visual impairment and estimates of retinopathy of prematurity at regional and global levels for 2010. Pediatr Res. 2013;74:35–49.
25. Gilbert C, Fielder A, Gordillo L, Quinn G, Semiglia R, Visintin P, et al. International NO-ROP Group. Characteristics of infants with severe retinopathy of prematurity in countries with low, moderate, and high levels of development: implications for screening programs. Pediatrics. 2005;115:518–25.
26. Kilangalanga NJ, Nsiangani LN, Dilu AA, Moanda KA, Ilunga MJ, Makwanga ME, et al. Epidemiology of childhood blindness and low vision in Kinshasa-democratic Republic of the Congo. Ophthalmic Epidemiol. 2020;27:45–51.
27. Gyawali R, Bhayal BK, Adhikary R, Shrestha A, Sah RP. Retrospective data on causes of childhood vision impairment in Eritrea. BMC Ophthalmol. 2017;17:209.
28. Zheng L, Do HH, Sandercoe T, Jamieson RV, Grigg JR. Changing patterns in paediatric optic atrophy aetiology: 1979 to 2015. Clin Exp Ophthalmol. 2016;44:574–81.
29. Muhit M, Karim T, Islam J, Hardianto D, Muhiddin HS, Purwanta SA, et al. The epidemiology of childhood blindness and severe Preterm-associated visual impairment and estimates of retinopathy of prematurity at regional and global levels for 2010 in Indonesia. Br J Ophthalmol. 2018;102:1543–9.
30. Zepeda-Romero LC, Barrera-de-Leon JC, Camacho-Choza C, Bernal CG, Garcia EC, Diaz-Alatorre C, et al. Retinopathy of prematurity as a major cause of severe visual impairment and blindness in children in schools for the blind in Guadalajara city, Mexico. Br J Ophthalmol. 2011;95:1502–5.
31. Haddad MA, Sei M, Sampaio MW, Kara-Jose N. Causes of visual impairment in children: a study of 3,210 cases. J Pediatr Ophthalmol Strabismus. 2007;44:232–40.
32. Patel DK, Tajunisah I, Gilbert C, Subrayan V. Childhood blindness and severe visual impairment in Malaysia: a nationwide study. Eye. 2011;25:436–42.
33. Hornby SJ, Xiao Y, Gilbert CE, Foster A, Wang X, Liang X, et al. Causes of childhood blindness in the People's Republic of China: results from 1131 blind school students in 18 provinces. Br J Ophthalmol. 1999;83:929–32.
34. Tsai LT, Hsu JL, Wu CT, Chen CC, Su YC. A new visual stimulation program for improving visual acuity in children with visual impairment: a pilot study. Front Hum Neurosci. 2016;10:157.