

ESLRR symposium

Cerebral visual impairment (CVI) in children from an interdisciplinary perspective



Chair: Susanne Trauzettel-Klosinski, MD, PhD

Co-Chair: Christiaan Geldof, PhD

After professional training in ophthalmology, Dr. Susanne Trauzettel-Klosinski worked in neuro-ophthalmology as Assistant Professor, Senior Physician and consultant at the University of Tübingen, Germany and Bern, Switzerland for 12 years. She did her “Habilitation” in Ophthalmology in 1994 and became Professor of Ophthalmology in 2000. In 1991 she founded and established the Low Vision Clinic and Research Laboratory at the University Eye Hospital in Tübingen and was the Director of this Institution until 2013. Since 2013 she is Head of the Vision Rehabilitation Research Unit.

All these activities have been dedicated to build the bridge between neuro-ophthalmology and vision rehabilitation. She has intensive international activities, multiple interdisciplinary international co-operations and functions in international societies, such as: vice president of ISLRR (2002-08), vice president of ESLRR (since 2010), Research Director of the European Neuro-ophthalmology Society (EUNOS), Fellow in the North American Neuro-ophthalmology Society (NANOS), coordinator of a European Research Project on reading in AMD. Her main research fields are: neuro-ophthalmology and low vision, reading disorders (developmental and acquired) and rehabilitation of patients with visual field defects (e.g. due to hereditary retinal disease, age-related macular degeneration, stroke), development of standardized tests and specific training programs.

Symposium outline

CVI is the leading cause of visual loss in children in the developed world, mainly because of the improved survival rates of premature infants. The exact definition and diagnosis of the condition are still a challenge. Cognitive dysfunctions are an added problem. Diagnosis affords subtle and specific tests for a wide range of visual disabilities from severe vision loss to dysfunction of perception and visuomotor integration. The topic will be discussed from a neuro-pediatric perspective presenting valuable screening tools as well as considering children with cerebral palsy. The special conditions of very preterm born children will be presented from healthcare psychology perspective. The role of attention for rehabilitation of

homonymous field defects focuses on the fact that patients can direct their gaze to a blind area of the visual field by consciously directing sustained focal attention there. The neuro-ophthalmological perspective demonstrates the effectiveness of a children-adapted explorative saccadic training to improve gaze strategies and quality of life. The educational perspective presents new task-dependent approaches, which help to observe and assess strategies for everyday life and in educational settings.

Symposium speakers

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| 1. Jenefer Sargent, UK | Cerebral visual impairment: a neuro-pediatric perspective |
| 2. Elisa Fazzi, Italy | Cerebral visual impairment in children with cerebral palsy |
| 3. Christiaan Geldof, NL | Cerebral visual impairment in very preterm born children: visual and attention functioning |
| 4. Manfred Mackeben, USA | Looking at what you cannot see: the role of attention |
| 5. Susanne Trauzettel, Germany | CVI with hemianopia in children: rehabilitation from a neuro-ophthalmological perspective |
| 6. Renate Walthes, Germany | Children with cerebral visual impairment: an educational perspective |

ABSTRACTS

1. Cerebral Visual Impairment: a neuro-paediatric perspective

Jenefer Sargent

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Epidemiological studies investigating the incidence and causes of severe visual impairment in childhood indicate that brain related visual impairment, or CVI, is increasing in importance in the industrialised world. However, such studies often rely on case notification by Ophthalmologists using a key feature of 'poor visual responses not due to known ocular pathology'. This presents a challenge since poor visual responsiveness in infancy and childhood may occur due to both visual and non-visual factors, and studies rarely report a full range of characteristics for each individual child deemed to have CVI. What exactly is being reported in such studies therefore may be less clear than the statistics apparently convey.

Nevertheless, it is likely that children who have brain pathology likely to interrupt dorsal or ventral stream pathways, are at risk of limitations on their visual performance. However, it

may not be clear how or even when such impairments may manifest, since the consequences of early brain injury may not be manifest until particular developmental stages have been reached.

One challenge therefore facing clinicians and researchers interested in childhood CVI, is the current lack of agreement on how the condition should be defined, how it manifests and which diagnostic tools (history and assessment) are required to distinguish it from other comorbid developmental conditions.

The emergence of screening tools for identification of possible CVI may lead to premature conclusions if full evaluation for other comorbid conditions such as cognitive impairment, attention deficits and social weaknesses does not follow. The paediatrician with expertise in developmental disorders is well placed to help evaluate the 'whole child' and to identify all areas of difficulty.

2. CEREBRAL VISUAL IMPAIRMENT IN CHILDREN WITH CEREBRAL PALSY

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Study objectives: children with cerebral palsy (CP) present a wide spectrum of visual disorders which include both peripheral problems and cerebral visual impairment (CVI), a deficit of visual function due to a malfunctioning of retrogeniculate visual pathways in absence of any major ocular disease. The aim of our study is to describe the neurovisual profile in children with spastic CP at school age, define the different CVI profile according to the types of CP and to explore brain structural alterations using Diffusion Tensor Imaging (DTI).

Methods: Thirty-one CP were selected from those consecutively referred at our Department during the last three years, according to the following criteria: spastic CP documented by neurological examination and brain MRI, school age, verbal IQ levels > 70, normal/near normal visual acuity. All the subjects were submitted to an evaluation of neurovisual functions with particular regard to ophthalmological, oculomotor, perceptual and visuocognitive aspects. DTI analysis were performed in nine CP patients and in 13 age-matched healthy controls.

Results: this study underlines the high risk of visual disorder in CP children (97% of cases). Subjects with bilateral CP present a more extensive visual involvement than hemiplegic

children. DTI analysis documented that CP children with visual-cognitive dysfunction presented lower fractional anisotropy levels in superior longitudinal fascicle (SLF) that is the principal fiber bundle related to cognitive visual function.

Conclusions: neurovisual dysfunctions are an integral part of the clinical picture of CP. A complete neurovisual evaluation appears to be essential in this clinical population in order to identify ocular, oculomotor, perceptual and/or visuo-cognitive dysfunctions early and to apply specific habilitation programs. The SLF lower fractional anisotropy levels observed in children with CP and visual-cognitive dysfunction confirms that SLF is a central pathway for visuo-cognitive abilities.

3. Visual and attention functioning and cerebral visual impairment (CVI) in very preterm born children.

Christiaan Geldof

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Very preterm birth (<32 weeks of gestation) is a risk factor for adverse ocular as well as cerebral development and subsequent visual and neurocognitive dysfunctions. This study aimed to characterise these dysfunctions and to study the interrelations between oculomotor, visual sensory, visual perceptive and visual attention functioning in 5 year old very preterm children and term born controls. Additional aims were to classify cerebral visual impairments (CVI) using empirical criteria and study the associations between CVI and intellectual and behavioural functioning.

A specific pattern of dysfunctions was found in very preterm born children, including impaired ocular alignment, visual acuity, visual field, stereovision, visual spatial perception, visual search and executive attention. Visual deficits that were classified as cerebral visual impairment (CVI) were associated with a range of behavioural problems including social functioning, and with dysfunctions in visual search and performance intelligence.

Very preterm birth is a risk factor for visual dysfunctions, also when ocular pathology is absent. Visual dysfunctions that were classified as CVI form a heterogenic picture in terms of number and severity and are associated with a wide range of neurocognitive and behavioural dysfunctions. Therefore, CVI in very preterm born children seems a marker of adverse brain

development and subsequent neurodevelopmental difficulties, thereby challenging the validity of the concept of CVI and stressing the need for differential diagnostic criteria.

4. Looking at what you cannot see – the role of attention

Manfred MacKeben

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Scanning eye movements involve transient attention, which is reflex-like and quick. However, it also requires a visible stimulus in the environment, which makes it an “exogenous” mechanism. For patients with a blind area in the visual field (hemianopia, peripheral field restriction) it is desirable that they perform eye movements into those areas to enlarge their field of gaze, but there is no visible stimulus there. These patients have another mechanism available by using sustained attention (SA), which is an “endogenous” mechanism that needs no external stimulus but can be directed at a location in space. SA can be controlled by volition and is slow.

Willful deployment of SA can trigger an eye movement that brings a previously unseen object onto the fovea. Research has shown that this form of gaze control can be trained to become a standard tool to augment orientation and mobility. This way, the field of gaze is extended and can contribute to conscious perception of the world surrounding us. Patients with homonymous hemianopia can learn to drive a car again, at least under restricted conditions. Their new gaze strategy is relatively simple, because they only have to learn to make saccades in the direction of the blind hemifield.

On the other hand, patients with advanced glaucoma can suffer from “tunnel vision”. They can improve their mobility by learning how to avoid bumping into objects. Their strategy needs to be more complicated than in hemianopia, because the extension of the visual field can lie in any direction. In both cases, it is noteworthy that deployment of SA does not require a physical stimulus, but only a location in the visual field.

SA has been researched extensively in humans and in primates. Studies have indicated that activating SA correlates with neural activity in the parietal and frontal cortices.

5. Cerebral visual impairment with hemianopia in children: rehabilitation from a neuro-ophthalmological perspective

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Background: Cerebral visual impairment is often associated with lesions of the suprachiasmatic visual pathways causing hemianopic field defects (HFD) with difficulties in spatial orientation. Rehabilitation from a neuro-ophthalmological perspective focusses on compensating strategies. The present study investigates, whether a children-adapted explorative saccade training (EST) can improve their eye movement strategies and performance in daily life, which we had shown in adult HFD- patients in a previous randomized, controlled study (Roth et al 2009).

Participants: 22 children with HFD (median age 11 years, 8 months (6-19y)) and 16 healthy normally-sighted age-matched children (median 11y6m, 7-17y) were included.

Methods: Visual fields were examined by different techniques. EST with a computer-based search task was developed for children. The HFD-children trained at home 15 minutes/day, 5 days/week, for 6 weeks. Search times (STs) during different search tasks were assessed before training (T1), directly after training (T2) and 6 weeks after end of training (T3). Gaze strategies were recorded by eye tracker. Control subjects performed the task at T1 and T2 (without training) to determine the effect of just repeating the search task. Standardized quality of life questionnaires (QoL) for children and specific orientation questions were evaluated.

Results: In patients, STs decreased in all search tasks from T1 to T2 and were sustained at T3. Eye movement strategy became more effective. QoL –scores improved. No changes occurred in the control group.

Conclusions: Children with HFD benefited from EST, indicated by shortened STs and more effective gaze strategy after training, which was maintained at T3, showing that the newly learnt gaze strategy could be applied to everyday life. Parents and children reported subjective improvement in daily life.

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6. Children with CVI from an educational perspective

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From an educational perspective the impairment itself, regardless of whether it is ocular or cerebral in nature, is not the critical point for initiating assessment and intervention. However observing the child's activity and its's limitations in participation guides assessment. Children born with visual processing problems are usually unable to tell how their vision differs from the vision of other people. Therefore, it is crucial to develop task-dependent approaches which help to observe and assess strategies for everyday life, at home, at kindergarten and at school. Indeed, children with suspected visual processing problems show a broad variety of behavior in order to master social and educational requirements. Pro-VisIoN (Processing visual information in Children) studies visual abilities as well as the child's strategies in responding to the demands of a task using multiple methods, including conversations with the child and family, observation of child strategies including eye-tracking, standardized tests such as visual acuity and documentation of oculomotor functions. Based on the visual profile of Hyvärinen and Jacob (2011) we will present first results of the assessment and follow up of 200 children.