General discussion

The overall aim of this study, supported by Royal Dutch Visio, Centre of Expertise for blind and partially sighted people, was to provide new information to advance efficient evidence-based diagnostic procedures for cerebral visual impairment in children, in order to improve detection and neuropsychological diagnosis in clinical practice.

We first investigated the potential of the crowding ratio for early detection of cerebral visual impairment on different levels of the care chain. Next, we specifically focussed on aspects of dorsal stream dysfunction (motion perception, object recognition, visual attention, visuoconstructive functioning), investigating the applicability and shortcomings of available quantitative neuropsychological tests for their diagnosis and detection. We further performed first epidemiological explorations of dorsal stream dysfunctioning in children.

Because neuropsychological testing is only reliably feasible from the age of 4 years onwards, we included children aged 4 to 7 years into the study, both children at risk for cerebral visual impairment because of (suspected) brain damage and a control group of typically developing school children.

For younger children and children with disabilities, a joint collaboration with the Neuroscience department was initiated for the development of a new diagnostic method based on the assessment of (reflexive) eye movements towards visual stimuli using remote eye tracking. Here, we performed exploratory data analysis on normal gaze patterns to motion coherent stimuli and to items that require the ability to visually integrate parts of the representation of objects into a whole (Gestalt perception) by quantification of reaction times and fixation qualities.

PRINCIPAL FINDINGS

- The crowding ratio can be used as an easily applicable detection method with sufficient sensitivity and specificity to distinguish children at risk for cerebral visual dysfunctions (children with brain pathology) from children with ocular pathology only, in groups that are referred for ophthalmological or specialised low vision assessment.
- However, the presence of crowding cannot be used to directly screen for cerebral visual dysfunction, because our analysis indicates that abnormal crowding is an independent impairment, which can co-occur with perceptual problems, especially in children with brain damage.

- For screening in the unselected children's population, as in youth health-care and paediatrics, the currently applied crowded visual acuity remains the best diagnostic measure.
- Normal limits for motion perception tasks can be based on chronological age in typically developing children, but in children with (suspected) brain damage, results should be evaluated against performance or developmental age. Based on chronological age, motion perception problems are likely to be overestimated in this group.
- Because a bottom effect was found for the biological motion task, this task is considered too difficult for children with a developmental age of 4 to 7 years. The remarks of several children that the phase-scrambled point-light figure looked like a dancing person, suggests that top-down processes like expectations might have influenced the results and makes the construct validity of the task questionable.
- In children with (suspected) brain damage, motion perception problems are specifically found for global motion and motion-defined form, but less often than has been reported for very low birth-weight children[1] and prematurely born children born preterm and with mild periventricular brain damage.[2] Motion speed problems are scarce in this group: we hardly found any abnormalities. This is partially due to missing data as a result of problems with the long total test time.
- Object recognition problems are associated with weak motion perception and visual attention, but not with weak visuoconstructive functioning.
- Dorsal stream problems (motion perception, object recognition, visual attention, visuoconstructive functioning) are rather specific and heterogeneous than general. We conclude that, if in individual children a dorsal stream function is found to be impaired, it cannot be predicted whether and which other dorsal stream aspects may be affected, too.
- Quantitative assessment of orienting responses towards visual stimuli using remote eye tracking is finding its acceptance in Dutch low vision centres.[3] The age dependent norm references presented in this study enables the quick and easy objective identification of motion perception problems in children at all age groups.
- The performances on Gestalt perception are weak in children with visual impairments. Specifically, children with brain damage had poor abilities in recognising the animate items compared to the children without brain damage. The eye tracking results add valuable data to grade the performance in terms of effort and task efficiency.

Improvement of clinical practice

To what extent can findings of this study already be implemented in clinical practice of expert low vision services and the chain of care suppliers before them? Our research mainly focussed on the diagnostic process within specialised low vision services, assuming that children at risk for cerebral visual impairment are referred. Among Dutch professional disciplines involved in the care supply chain (youth healthcare specialists, paediatricians, paediatric neurologists, rehabilitation specialists, (paediatric) ophthalmologists, general practitioners, intellectual disability physicians, orthoptists, optometrists, opticians), different guidelines for the detection of visual problems are in use. Most of these focus on the detection of ocular abnormalities, impairment of visual acuity, strabismus and amblyopia (Table in Appendix),[4-10] but cerebral visual impairments are mentioned in the guidelines for youth healthcare specialists [6] and ophthalmologists.[9]

In the youth healthcare guideline, prematurely born children, children with cerebral palsy, children with hydrocephalus or perinatal asphyxia and children with intellectual disabilities are considered the primary risk groups.[6] However, the awareness of cerebral visual impairments is still relatively low among professionals involved in early detection and referral, as has already been pointed out in 2007 by a multidisciplinary Dutch expert working party on early detection of cerebral visual impairment.[11] It took until 2016 before the development of a Dutch multidisciplinary guideline, specifically aimed at cerebral visual impairment in children, was initiated. This has now (*September 2018*) resulted in a well-designed and complete draft guideline, nearly ready for publication, including all disciplines that are involved in the care chain from early detection to specialist low vision services.

Table 1 shows, that not only the heterogeneous aetiology and manifestations of cerebral visual dysfunctions and the lack of consciousness of professionals, but also other factors, such as a lack of valid screening instruments and diagnostic tests, limited resources, and current eligibility criteria for specialist low vision services, may impede timely detection, consistent referral, and early or long-term intervention or habilitation.

Table 1. Impeding factors for detection, habilitation and intervention of cerebral visual impairment

General practitioner, neonatologist, paediatrician, child neurologist, rehabilitation specialist, intellectual disability physician	**(Child) ophthalmologist, orthoptist, optometrist, optician**	**Special services for visually impaired people or low vision services**
o Unconsciousness of cerebral visual impairment, especially of impairments other than low vision o Lack of easily applicable detection instrument o Assumption that assessment of visual functions in children with an intellectual disability or developmental delay is - too difficult - too big a burden for the child - not useful in all children	o Limited resources to assess all children at risk, specifically children with developmental delay or intellectual disabilities o Unconsciousness of possible impairment of visual functions other than visual acuity and visual fields o No referral of children with (sub)normal vision for evaluation of visual perception	o Lack of objective eligibility criteria for neuropsychological assessment o Not all visual functions in children can be assessed with quantitative tests o Dutch eligibility criteria for long-term intervention and habilitation and for accessibility to and refunding of aids and appliances insufficiently include children with cerebral visual impairment and (sub)normal visual acuity

Recommendations for the care supply chain

So, it is of paramount importance that physicians in public youth healthcare services, general paediatrics, general practice and intellectual disability clinics timely identify children at risk for visual function problems and refer them for ophthalmological evaluation. Based on our findings, described in **Chapter 2**, we recommend that screening of visual acuity by youth healthcare professionals remains essentially the same as now. At school age, many children with moderate and severe disabilities are not able to cooperate with regular visual acuity testing methods. For such children, the guideline for screening of visual functioning by youth health physicians recommends the use of LH symbols.[7] If LH symbols are not applicable, assessment with the Teller Acuity Cards is suggested for young children with no or low verbal abilities but can be applied in school children with intellectual disability, too.

Members of the 2007 expert working party, mentioned above,[11] youth health physicians working in special schools for children with disabilities, paediatricians specialised in congenital and genetic conditions, and intellectual disability physicians, stressed the need of an easily applicable test to detect potential cerebral visual impairment. Therefore, in our opinion, addition of *crowding* to visual screening in special schools and intellectual disability clinics - where the risk groups are found - is worth considering, although its application is restricted to children with mild intellectual disabilities. An evaluation of its effectivity would be an interesting research project. If effective, this may decrease the burden on ophthalmology clinics and low vison care.

In the Netherlands, young children in at risk groups who are checked by paediatricians, such as children with severe intellectual and motor disabilities, may not visit regular maternal and child healthcare centres and do not participate in regular preschool vision screening. The same working party proposed a safety net construction for children in at risk groups, with routine orthoptic/ophthalmological referral by the vaccinating child healthcare professional or paediatrician at age 11 months and 4 years.[11]

In the Netherlands, eligibility requirements for referral to specialist low vision services, as formulated in the guideline of the Dutch Ophthalmological Society,[9] are followed for reimbursement decisions according to the Exceptional Medical Expenses Act or by regular healthcare insurances. This guideline has been updated in 2011. Whereas the former version focussed predominantly on visual acuity and visual fields,[12] in the updated guideline specific attention is asked for cerebral visual impairment in the introduction of chapter 7 on visual impairment in children.[9] It is remarked that cerebral visual impairment is insufficiently recognised in clinical practice, among others by late or no referral by ophthalmologists to low vision rehabilitation centres. However, no other referral criteria are given than the outline of risk groups with 'mental retardation', low birth weight, and cerebral paresis. This lack of clear-cut criteria is confusing and may harm the development of children with cerebral visual impairments and a normal visual acuity, by denying them essential aids or long-term expert care.

Therefore, we explicitly recommend addition of crowding assessment to standard paediatric ophthalmologic and orthoptic assessments of children with intellectual disabilities that are able to match or name symbols, because it would partially solve the existing eligibility problem. Addition of crowding assessment to routine diagnostics may help decide which children at risk should be referred for expert assessment of low vision. According to the current eligibility criterion for Dutch low vision services (visual acuity 0.5 or lower), only 13 out of our 26 study participants in which crowding was assessed, would have been referred by the ophthalmologist to a low vision service, whereas a crowding ratio of 2 or higher was found in 19 out of 26 patients.[13] Although these data are still limited, they indicate that referral might improve if based on the crowding ratio.

Professionals in ophthalmologic care know and accept the concept of crowding, whereas binocular assessment of crowded acuity is common routine. To implement and quantify crowding, an assessment of single optotype acuity has to be added, which would take a few additional minutes. In principle, a crowding ratio can also be assessed with other optotypes than the Cambridge

Crowding Cards. One should be aware that, because of differences in inter-symbol spacing and step size (for example 1/3 or 1/2 octave) that define the letter size of consecutive lines, the norm value for crowding may be different between tests. Nevertheless, a ratio of 2 or more always remains an alarm sign.

Recommendations for eligibility criteria for low vision services

Not all children with cerebral visual impairments will be identified by crowding assessment, whereas it will take time until crowding assessment has been completely implemented in ophthalmologic routine. So, apart from our recommendation to add crowding to standard paediatric orthoptic assessments, addition of 'confirmed brain damage' or 'explicit risk of brain damage' to the eligibility criteria will be a large step forward. Indeed, since the update of the ophthalmology guideline in 2011,[9] children with (a risk of) brain damage are now regularly referred by ophthalmologists; this should become standard in case of doubt about the child's visual perceptual functioning.

Epidemiological research into prevalences as well as consequences of cerebral visual dysfunctions in children at risk is urgently needed to support this claim.

Recommendations for expert assessment of cerebral visual functions in low vision centres

Crowding

In low vision services, the decision which children to select for neuropsychological assessment of cerebral visual functioning would be facilitated by adding crowding assessment to routine orthoptic intake assessments. In recent years this has already become more common.

Professionals should be aware that results are likely affected by test characteristics, like symbol spacing (fixed or proportional) and test distance, and patient characteristics, i.e. presence of absence of brain damage, low vision or nystagmus.[13-15] Results of the study of Huurneman et al[15] suggest that testing at a long distance (5 m) with a test with a fixed symbol spacing results in higher crowding in children with low vision and nystagmus. Children with low vision but no nystagmus performed comparable to children with normal vision. In our study with a chart with a proportional symbol spacing (inter symbol spacing 50%)[13] children with ocular abnormalities, including nystagmus, performed comparable to children without abnormalities, whereas crowding was higher in children with brain damage. For children with cerebral visual

impairment, testing at a distance may be more difficult than for children with ocular abnormalities, leading to worse outcomes. This could be investigated, too, in the research project in youth healthcare, suggested above.

Crowding problems may also have implications for reading.[16] If all crowding ratios are size independent, as in amblyopics, critical spacing assessed with the crowding cards can be used to estimate critical spacing for reading (spacing between letters).[16] Adjustment of the spacing in text according to the individual's critical spacing could then optimise reading rate. This could explain why magnification is generally considered to help, because spacing as well as letter size increase.

Neuropsychological assessment

Based on **Chapter 6 and 7** we recommend that multiple aspects and functions should be routinely included in neuropsychological assessment of dorsal stream dysfunction in children at risk, because problems were found to be mostly isolated and heterogeneous. In low vision services, use of the child´s performance age in the interpretation of test outcomes, as recommended and used in **Chapter 5-7**, is increasing, but not yet standard. We showed in **Chapter 6** how the use of chronological age leads to overdiagnosis of motion perception dysfunction.[17] Testing of performal IQ or expert estimation of performance age should be routinely performed by psychologists and behavioural scientists in low vision services. Efficiency of different diagnostic strategies should be systematically evaluated.

Generally, the neuropsychologic tests applied in this study appeared valid and applicable, but several aspects can be improved. On a group level, they discriminate between children at risk and typically developing children. However, normal limits remain a bottleneck for all tasks, requiring evaluation in larger samples. This does not necessarily hamper their current clinical application as observational instruments: confidence intervals of normal limits may give an indication whether a child belongs to the weakest 5 or 10 percent performers, but more work has to be done to obtain valid reference values.

For subtests of the L94 in which children have to name objects (VIEW, De Vos, NOISE), the influence of children's verbal development and word frequency used in their socioeconomic environment on concurrent validity has to be investigated further, too. Current norms of the nearly 25 years old L94 should urgently be updated in new study groups.

In Dutch low vision services, quantitative neuropsychological tests of motion perception and visual search or selective visual attention, as applied and evaluated in our study, are not yet part of routine neuropsychological

diagnostics. At best, unspecified observational assessment of visual following can be part of orthoptic assessments, whereas motion perception dysfunction may be considered if the child has problems in traffic situations. In such cases of suggestive problems in daily life, it may be relevant to include specific tests of motion perception in the assessment.

Finally, we recommend some technical improvements to the tests. A touch screen would facilitate the application of computerised object recognition, visual search and motion perception tasks, because that would eliminate the necessity of an observer response. Several adaptations would improve the visual search task: 1. a button as a starting point because the children had to be urged to put their hands on the table before each trial, 2. video registration to observe typical behaviour, and 3. addition of a short response inhibition test.

Applicability of available and new diagnostic tests

For large-scale epidemiological research of cerebral visual impairment (prevalence, risk groups, disability, natural course), valid, precise and not too time-consuming measures are needed, with well-evaluated reference values. Currently, only samples with limited sizes and selected pathology have been evaluated. To obtain large enough study populations, international collaboration of research groups will be necessary. To that end, international consensus should be reached on a limited battery of diagnostic tests to further evaluate. In the discussion paragraph of our systematic review (**Chapter 4**), we have made a first, argued selection on a basis of technical specifications as well as psychometric properties.

We also recommend cross-sectional as well as longitudinal research into the connection of outcomes of diagnostic tests and visual problems experienced in daily life. The crowding ratio can be used for epidemiological research into the prevalence of crowding, its natural development during early childhood, and if persistent, its relationship with brain damage and amblyopia, as a basis for its validity as an easily applicable screening method.

After further studies of its validity, the remote eye tracker method might enable objective assessment of aspects of visual functioning in very young and intellectually disabled children and might provide additive information that helps to interpret classical behavioural data.

Quantitative assessment of orienting responses towards visual stimuli using remote eye tracking may improve detection and diagnosis of visual function problems, specifically in very young and difficult-to-test children.[18-21]. From 2010 onwards, studies were published on the quantification of (reflexive)

orienting responses in terms of Reaction Time to Fixation (RTF), fixation durations and fixation accuracies[22, 23] resulting from visually-guided orienting responses. Here, eye movements were introduced as a read out of brain functioning, i.e. bottom-up driven processes. Application within a large group of healthy children between 1-12 year of age not only showed age-dependence of orienting responses but more importantly, differences in RTF's between different visual stimuli.[24] It was suggested that RTF represents the efficiency of processing specific visual features, such as contrast, colour, form coherence and, most importantly, motion coherence. Indeed, in **Chapter 8** we showed that with this approach, age-related processing of three different types of coherent motion could be identified in a group of typically developing children aged 1-12 years.[25] Thus, identification of prolonged reaction times and fixation pattern abnormalities are potential screening markers. Its further development, including longitudinal evaluations,[26] has recently led to implementation of eye tracking within low vision centres, an initiative between the Neuroscience department of Erasmus MC and Royal Dutch Visio.[27]

Recently, an increasing number of studies have investigated the advantages of eye tracking based methods to test amongst others visual acuity and visual field defects as part of visual function assessments. Still, a very limited number of studies are available that combine eye tracking with visual perception tests. Whereas bottom-up processes are stimulus-driven, top-down processes are related to inner motivation and cognitive cues.[28] In **Chapter 9**, we presented one of the first studies on Gestalt perception in visually impaired children. Here, the emphasis was not on processing speed or accuracy, but on fixation qualities as a measure for (sustained) visual attention. Our study not only suggests that children with visual impairments, independent of the cause, perform worse on Gestalt perception, but it also shows that the performances of children with CVI are weaker than that of children with OVI. Children with CVI perform in particular weak on the animal items. Eye tracking results suggest that differences between patients groups might be explained by different strategies: children with OVI that perform weak tended to fixate shorter, whereas children with CVI need in total more time to perceive and respond. Did children with OVI put in less effort than the children with CVI? The addition of an additional eye tracker parameter, i.e. pupil diameter, might help to solve this question, because pupil diameter is positively related to the workload: i.e. the pupil diameter is larger in tasks with a higher workload.[29] Studying pupil diameter might also clarify whether animal items are really more difficult than object items. And does it predict performances in general and at item level?

We conclude that our study has considerably added to insights into the value and shortcomings of internationally available diagnostic tests for dorsal stream function problems in children, leading to specific, underpinned recommendations for selection of tests and further steps in research, as well as technical improvements of the tests, used in the current study. The eye tracker paradigms introduced in this thesis even enable quantitative testing of motion perception problems in very young and intellectually disabled children and Gestalt perception in children suspected of brain damage. This new approach will definitely add value to the existing neuropsychological assessments.

We established that dorsal stream problems in children with (suspected) brain damage are rather isolated than generalised, so multiple aspects should be part of neuropsychological assessments. And we evaluated crowding as a screening method for cerebral visual dysfunction. Although a lot of work has yet to be done before the tests are completely fit for use in clinical practice, we could formulate first recommendations for improvement of both the referral process towards specialised low vision services and within these services, the selection of clients for neuropsychological assessment.

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Appendix 1

Appendix 1. Care supply chain for children with visual impairments according to existing Dutch guidelines for different professional disciplines

<table>
<tr><th colspan="3">Pregnancy and birth
Complications?</th></tr>
<tr><th colspan="2">No</th><th>Yes</th></tr>
<tr><td colspan="2">General Practitioner or Midwife [1, 2]
Health screening, including inspection of the eyes
Referral to ophthalmologist within a week if:
o Suspicion of congenital ocular abnormalities, specifically cataract and glaucoma
Consultation or referral if:
o Conjunctivitis</td><td>Pediatrician, neonatologist or child neurologist
Developmental delay and brain damage:[3]
As part of diagnostic procedure: referral for orthoptic/ophthalmologic assessment including strabismus, slitlamp biomicroscopy, ophthalmoscopy

Down Syndrome:[4]
Referral for active orthoptic/ophthalmologic screening at different ages</td></tr>
<tr><td>Youth healthcare physician/ nurse [5]
Preschool and school age:
Scheduled screening of health and development, including inspection of the eyes and screening of distance visual acuity

Referral to ophthalmologist in case of:
- Fixation/following problem
- Abnormal ocular structures
- Monocular/binocular distance visual acuity < 5/10
- Suspicion of cerebral visual impairment (risk groups)</td><td>General Practitioner [6]
Assessment of distance visual acuity and refractive errors if parents or patient complain about vision
Referral to ophthalmologist if:
o Visual acuity ≤0.2
o Wearing a correction
- spherical >± 7 D or cylindrical >± 3 D
o Monocular distance visual acuity: ≤ 6 years >0.2 and <1.0 6-20 years >0.2 and <1.0 and myopia and/or astigmatism or other ocular abnormalities or unrecognised amblyopia</td><td>Down Syndrome Team [4]
Ophthalmologist or orthoptist in the team?</td></tr>
<tr><td colspan="3">Specialist assessment</td></tr>
<tr><td>Optometrist, Optician
Checks of ocular condition and refractive errors, prescription of glasses</td><td>Ophthalmologist and orthoptist [7]
Detection or exclusion of ocular abnormalities
Referral to low vision service if there is a request for help and one of the following criteria:
o Visual field defect:
- Concentrical <30°
- Hemianopsia
o Distance visual acuity:
- <0.3
- 0.3-0.5 + need special support
o Near visual acuity:
- < 0.25
- Insufficient reading with addition of +4
o Severe light sensitivity</td><td>Ophthalmologist and orthoptist in case of Down Syndrome:[4]
Active screening every 2 years, specifically including cataract, strabismus, amblyopia, refraction, nystagmus, visual acuity, accomodation, keratoconus</td></tr>
<tr><td></td><td>Special services for visually impaired people or low vision services
Diagnostics, treatment and habilitation</td><td></td></tr>
</table>

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