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RESEARCH ARTICLE

Approaching New Clinical Horizons: The Case of Cerebral Visual Impairment (CVI)

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ABSTRACT

Cerebral visual impairment (CVI) in children is, to our knowledge, a very common but until now very often neglected outcome of, among other things, a) a stressful pregnancy, b) preterm birth and c) stressful term birth with pre- and perinatal injuries. The entire visual system includes both the eyes and the brain, and damage can affect one or both of these organs. The visual brain-focused aim is that children gain precise visual access to their surroundings, linking visual information with the appropriate language and motor skills to both enable them to understand what they see and guide movements along the visual acuity. Thus, vision, language and motor skills typically evolve together during the first months of life.

The meaning of the so called 'seeing sense' is still not considered enough to understand its contribution to normal developmental stages and vision-related deterioration in childhood. A systematic diagnostic approach to CVI in the medical system is missing. Consequently, experimental interventions are very rare. However, due to the demographic development (e.g. the increase in preterm births) there is an increase in the prevalence and cerebral visual dysfunctions and the need for its treatment.

Early insult to the visual system affects more than visual brain perception. As visual brain processing is related to cognition, emotion, motivation and the motor system, early visual impairments could negatively influence the development of all these functional systems. Thus, human brain development depends on structural and functional visual conditions and can be significantly disturbed by cerebral visual impairment. This article emphasizes the influence of possible early damage to the visual pathways on general development and academic achievement and the implication of CVI in the development of affected children.

1. Introduction

Cerebral visual impairment (CVI) in children is, to our knowledge, a very common but until now very often neglected outcome of, among other things, a) a stressful pregnancy, b) preterm birth and c) stressful term birth with pre- and perinatal injuries[1,2]. Thus, the most common causes of childhood CVI typically occur while the child is in the womb, during birth or in early delivery when there is hypoxia[3-6]. Consequently, in these cases, the cerebral visual system is not sufficiently developed to address the visual challenges of the world as soon as the child is out of the womb. Brain malformations, cerebral haemorrhage, genetic defects, cerebral infections, traumatic brain injury and seizures are very often connected to CVI[6-8].

The entire visual system includes both the eyes and the brain, and one or both of these organs can be damaged. At birth, the visual system is like an empty library ready for different sets of books in different covers. This precedes programming of the visual system. *The visual brain* is a conceptual term referring to the brain elements that serve to support vision. The visual brain-focused aim is that children gain precise visual access to their surroundings, linking visual information with the appropriate language and motor skills to both enable them to understand what they see and guide movements along the visual acuity. Thus, vision, language and motor skills typically evolve together during the first months of life. However, evaluation of the developmental stages of early childhood is more focused on the language and motoric side because the visual brain contribution is rarely considered[1,9-11]. Developmental delay becomes more evident after six months of life, when the normal development of movement is learned progressively through search and exploration. When exploring the early interactions of the new-born child with their environment and early learning processes, the visual approach is widely neglected in the check-up examinations of the developing child, because the extent and meaning of visual brain development is not given enough attention by professionals due to a lack of measurable optical (eye) deviations. However, CVI is based on functional processing inside the brain rather than on structural landmarks or optical problems. Damage to visual centers of the brain is still not recognized as a significant cause of visual impairment. Neurologists are still convinced that seeing and understanding (recognizing and interpreting) are two separate functions. That means, either patients do not see, or patients are not able to label or identify what they are seeing. Debates still take place among medical and other professionals about the nature of CVI, and in many areas of paediatrics, the message is still: 'Visual processing and even more an impairment of this process does not exist.' (personal communication). Knowing the meaning of the so called 'seeing sense' is still not considered enough to understand its contribution to normal developmental stages and vision-related deterioration in childhood. A systematic diagnostic approach to CVI is lacking in the medical system[12]. Consequently, there are only very rare scientific evaluated interventions[9,13]. However, due to the demographic development there will be an increase in treatment requirements for cerebral visual dysfunction[14,15].

Early damage to the visual system does not only affect visual brain perception, which is related to cognition, emotion, motivation and the motor system, early visual impairments could negatively influence the development of all these functional systems. Thus, the development of the human brain is dependent on structural and functional visual conditions and can be significantly disturbed by a cerebral visual impairment. This article emphasizes the influence of possible early damage of the visual pathways on general development and academic achievement and the meaning of CVI to the development of affected children.

2. How the brain 'sees' and what are the somatic/psychological subsystems in reference to visual processing?

The photographic images of (people, fruits, books…) that are generated inside the eye must be converted into electrical signals. The information in these signals is then carried to the brain and processed. The brain creates the picture. Being able to recognize someone/something in a group or on a patterned surface requires us to have a picture 'library' in the brain that we can instantly compare with incoming data. The matching face/item is recognized, but all other faces/items are considered unknown. The library must be linked to predictive knowledge of the feature of the particular image and available for forward planning of our actions[16].

2.1 Visual functions

The visual cortex has different functional areas for the analysis of colour, form, movement and spatial information (e.g. position, distance and direction) and for objects, faces, places, text and number material. These functions can be differentiated in lower and higher visual functions.

2.1.1 Lower visual functions

The lower visual sensory functions are a) visual acuity, b) colour vision, c) contrast sensitivity, d) visual field and e) monocular and binocular depth perception. Additionally, the ocular motor functions (eye movement in harmony in all directions) belong to the lower visual functions.

2.1.2 Higher visual functions

Higher visual functions can be classified as a) visual perception, b) visual cognition, visual guidance of movement and d) visual attention. A child will not explore their environment in the absence of curiosity or without attention. Perception is fundamental to the development of attention, memory, executive functions, sensorimotor activation, language, motivation and emotion. In this context visual perception is the act of detecting and recognizing what is seen. Perception does not operate in isolation but is embedded in the network of higher mental functions. Visual cognition is considered as an extension of visual perception and involves the capacity to process what is seen, to think about its significance and to manipulate and use both, incoming image data and remembered imagery in the context of thought. This is called a mental action.

Visual guidance of movement refers to the mapping of incoming visual information, which is used to guide movement of the upper or lower limbs and the body. In other words, these are the visuo-motoric functions. The absence of visually guided reactions is related to the child's inability to look at and touch an object at the same time. These functionalities are not synchronized; the actions are obviously performed separately. Synchronization of this visuo-motoric processing also occurs in the action of writing and drawing. If motor and visual functions are not synchronized, the impression is given of an unreadable writing or painted pattern. Additionally, mechanisms in the brainstem, midbrain and thalamus are involved in the guidance and control of eye movements to fixate stationary and moving visual stimuli[17].

2.2 The visual brain system

The pathway of visual information from the retinal input starts in the nervus opticus, passes the optical chiasm and enters the brainstem, where the lateral corpus genicalatum (CGL, part of the thalami) is reorganizing the information according to the structural system of the retina[18] (Figure 1).

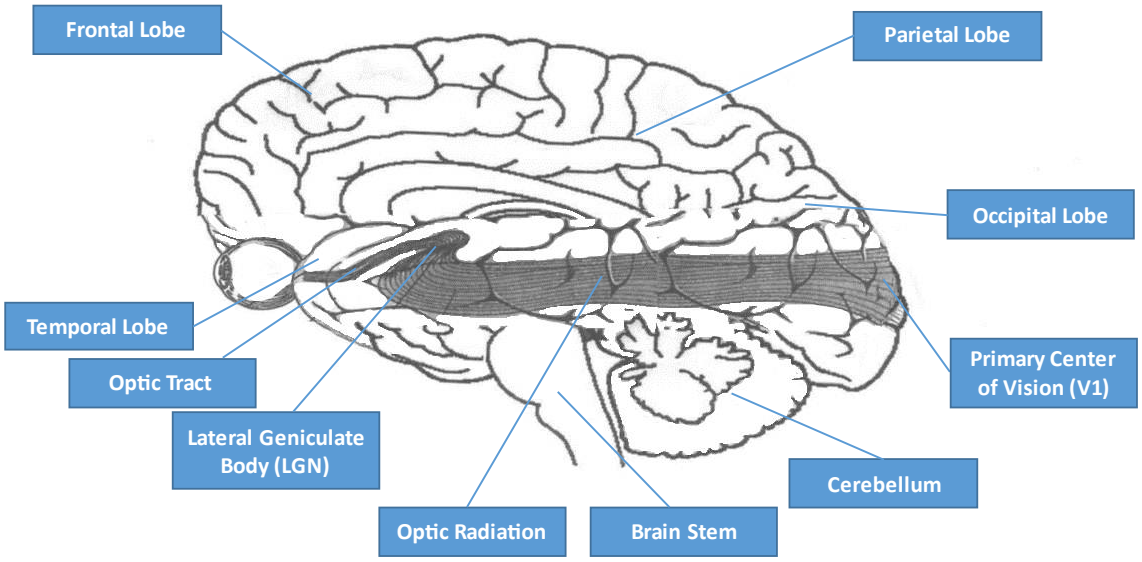

Figure 1: Visual pathways and the brain (basic figure source from Friederichs, P. and E.[19] with kind permission of Klett-Cotta publishing company and modified to Roman-Lantzy[10]

This means that although visual information inside the nervus opticus is mixed up, it will be sorted out inside the CGL according to a) the ganglion cell types inside the retina (magno- vs. parvocellular system), b) the left/right eye and c) the origin of the retinal position inside the fovea. Fibres of the

magnocellular ganglion cells coming from the peripheral retina are responsible for detecting and analysing moving images, while the parvocellular ganglion cells coming from the central area are responsible for detail analysis. These well-separated informational fibres will follow a path to the occipital lobe, where the original retinal structure will be projected. The retinal structure is more or less mirrored in the primary visual cortical field (V1) inside the occipital lobe, creating a visual map. Each point of the retina activates a corresponding field of neurons inside V1. Thus, the visual field projected to the occipital lobes can be seen as an improved version of information of the retina. Attention is brought to the visual system via the two thalami, which can be considered as relay stations where the structure of the retinal system is projected according to the specific eye and retinal position. In this context, retinotopia is challenged by the fact that ganglion cells on the retina from both eyes must be stimulated synchronously to realize a common receptive field for the same point in space. Neighbouring cells of the retina will be projected to neighbouring cells of the CGL and subsequently to V1, well differentiated by the eyes. This informational system is organized by parallel information processing in different transportation fibres, clear retinal topography, informational gating inside the thalami and multiple cortical representations. This process was investigated by the experiments of Hubel and Wiesel in the early 1970s[20,21]. As part of their work, Hubel and Wiesel demonstrated that the ability of cortical cells in the occipital lobes to interpret impulse messages from the retina develops in infancy and that even relatively short periods of light and pattern deviations can result in permanently impaired vision. They discovered the so-called 'eye dominance columns', demonstrating that specific cells inside the structure of the CGL and further on inside the cortical zones transport information from both eyes separately. The entire anatomy and physiology of the central visual process from the retina to the primary cortical visual area V1 is compatible with the idea that there are several different channels which will be processed in different parallel fibres to parallel inputs. Different types of fibres carry different visual information depending on their retinal location. Thus, the visual representation is not a projection like that of a camera. Each channel is specialized in analysing different aspects of visual perception.

Each thalamus can be considered as a sort of sensory filter. If a person decides to look at an object (or to listen to a sound), the thalamus opens up to allow visual (and auditory) information to flow to the cortical areas, resulting in further processing inside the different (dorsal and ventral) streams. At the thalamus side, the flow of visual information to the cortical visual pathways is modulated by altering the amount of attention paid to the visual modality (visual gating). Information from the visual sense is gathered and may then be processed or not processed to the cortical areas, depending on whether attention is given to the particular sensory system. Such information is analysed and synthesized in different cell assemblies inside the thalami. Visual processing requires a strict separation inside the thalami, a primary integration of signals in the occipital lobes and further specialized informational processing in the ventral and dorsal streams (figure 2). If the thalami are 'closed' (e.g. by auditory interferences or by suppression/interferences of unsynchronized visual processes), they prevent further visual processing from taking place[22,23].

Another visual processing feature (approximately 10% of the entire visual pathway) is brought through the superior colliculi and projected to the thalami. The superior colliculi on top of the brainstem affect thalamic gating by reacting reflexively to potentially hazardous visually stimuli in the outside world. This primitive pathway is already active soon after birth for the subconscious selection of gaze and is used by the new-born infant up to the age of three months[24] before the main partly conscious visual pathways connected to the prefrontal cortex are developed.

In the beginning, the new-born infant is only able to focus on one thing at a time but develops its visual system and learns to split attention by using their developing ventral and dorsal stream capacities. The aim is that children gain visual access to their surroundings by linking visual information obtained by thalamic gating with affiliated language to understand what is seen. However, it is always our primitive visual brain that helps to protect us further in our life. It detects a peripheral movement and automatically initiates a protective movement before we are aware of it. This pathway is responsible for the accompanying eye movements (saccades) directed at a spatial location, which are controlled by the prefrontal cortex in the frontal eye fields.

2.3. Beyond the primary area: What happens after the primary visual centre V1?

Visual processing is triggered by the outside world and guided by the inside one. The initial steps of visual processing occur in the occipital lobes as visual

representation of the outside world coming from the retina. This is realized by more than 10^9 cortical neurons, whereas the number of retinal ganglion cells is only in the order of 10^6. The size of the visually selected areas will be further determined by processes taking place inside the occipital, temporal and posterior parietal lobes and the brainstem (thalamus). It takes 100 ms for information from the retina to reach the occipital lobe. The occipital lobe sorts the information into component parts (contrast, colour perception, stereopsis, visual field [what can be seen] and the orientation and dimension of the image elements). Fundamental image processing takes place at this location. The left occipital lobe 'sees' the right sight of the visual scene and vice versa. The lower occipital lobe 'sees' the upper part and vice versa.

During the next 100 ms, the visual information will be analysed in two separate pathways by two main cerebral areas: a) the temporal lobes which are called the 'ventral stream' and b) the posterior parietal lobes, which are called the 'dorsal stream' (Figure 2).

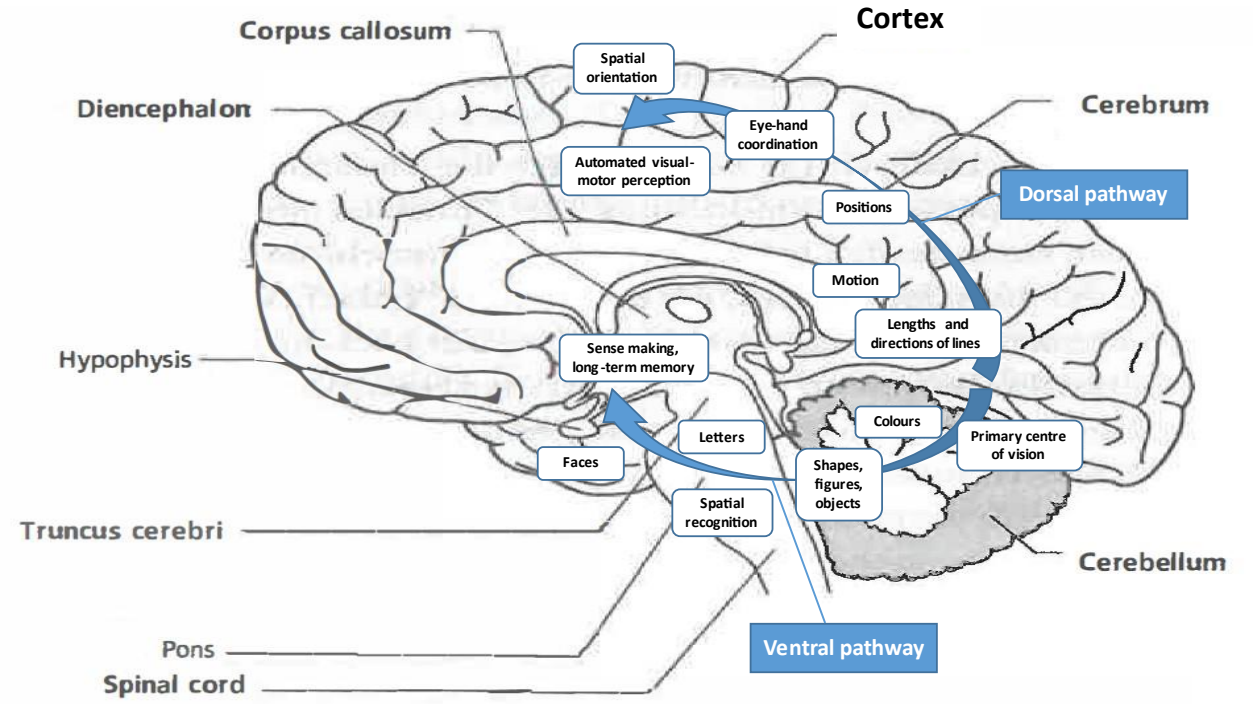

Figure 2: Cerebral visual processing with the dorsal and ventral stream (basic source from Friederichs, P. and E.[19] with kind permission of Klett-Cotta publishing company)

The ventral stream supports visual recognition by answering questions about the identity of objects, subjects and routes. It can be considered as an image store[25-27]. This is considered as a conscious operation. The dorsal stream is responsible for visual guidance of movement, visual searching and the switch from one element to another (simultaneous processing). It provides the awareness necessary for movements in terms of having an idea of directions and distances, body awareness and body orientation in space, including eye-hand orientation. Affective stimuli are processed along the dorsal pathway as well. All these processes function subconsciously inside the dorsal stream.

Both streams are closely connected by additional pathways and other perceptual systems. The ventral stream shows prolonged development and appears less plastic than the dorsal stream. Fibre connections within the ventral stream mature earlier than those in the dorsal stream.

Another important visual system was discovered by Cattaneo and Rizzolatti[28] and is called the 'mirror neuron system'. It enables the rapid awareness of other people's hand movements, gestures and moods. It mirrors our own feelings and motor functions. There is not much knowledge of this system at present, and a better understanding of it will require further investigation. The mirror neuron system competes with visual attention. It helps in understanding the action of others. It is a learning system in terms of observing the actions of others and copying them appropriately and in context.

3. What is cerebral visual impairment (CVI)?

The involvement of changes in visual pathways can be ocular, oculomotor, perceptual and cognitive. Cerebral visual impairment refers to disordered vision and/or visual perception of any type or severity as a result of damage or disorder to visual pathways or centres of the brain behind the chiasm and behind the corpus geniculatum laterale (CGL) inside the thalamus[1,18]. In the case of a CVI the eye 'sees' (e.g. detects visual stimuli), but the brain cannot make sense of visual information. Thus, cerebral visual impairment must be considered as a neurological disorder, which means that it refers to a brain condition rather than an eye condition. It must be kept in mind that cortical visual impairment is different from cerebral visual impairment[11]. Cortical visual impairment is associated with medical and educational impairment. Patients with this condition receive vision support services from specific teachers for students with visual impairment.

Chong and Dai[6] calculated from their data a CVI prevalence of 0.02% in the entire population of New Zeeland. Another study in the US showed a prevalence of 23.6% in visually impaired children[29]. This was confirmed by Boonstra et al. (2012), who found a prevalence of 27.2% in visually impaired children in a study in the Netherlands. A study with 46 preterm primary school students in Scotland revealed a 33% prevalence of cerebral-visual dysfunction[31].

3.1 Clinical effects of CVI

Vision is helpful but not necessarily essential for understanding space, spatial relationships and internal spatial representations. Behaviours exhibited by some children with visual impairment can be difficult for sighted children or adults to interpret because such behaviours may serve different purposes in visually impaired and sighted children. In general, 26% to 57% of patients with visual impairment suffer from emotional problems[32,33]. Developmental delays in visually impaired children can already be observed in the age range from 10 to 12 months[34]. If vision is impaired, children may gain visual understanding through the use of compensatory alternative sensory channels such as hearing and touching. Individuals with CVI can thus only interpret the visual world with the help of specific haptic adaptors and/or audible instructions. If these alternatives are unavailable or rarely available, children (e.g. those who suffer from CVI) whose hands remain at shoulder height for much of the time during the first months of life and who do not show evidence of exploratory tactile function may be at a greater risk for delayed development of object permanence and image formation[1].

Symptoms are often unusual manifestations of which patients complain. This requires awareness and knowledge that these manifestations might be related to visual impairments. If visual limitations are not recognized, affected children may be at risk of receiving incorrect therapies. For example, if a child does not see and becomes anxious because of unreliable images, general anxiety therapy will not work. Effects of CVI on development can vary enormously. Profiles can vary as a result of environmental interference and brain neuroplasticity, which can induce a reorganisation of maturing visual functions. They impact developmental stages later in life and the experience of the world in diverse ways[33].

Children´s unusual behavioural responses that result from visual disorders can be categorized. There may be a lack of visual communication response in the case of prosopagnosia/prosopdysnosia, a difficulty detecting someone's face and facial expressions. Such responses—lack of anticipation, joint attention and eye contact—may be misinterpreted as features of autism spectrum disorder. Further on, reactive and adaptive responses due to simultaneous agnosia, optic ataxia or dyskinetopsia can be observed. It may occur that children experience a visual world in which people or objects appear and disappear and do not obtain a mental explanation for their appearance or disappearance. In these cases, children can express angry outbursts, anxiety or panic attacks. These responses occur as compensation and may be misinterpreted as oppositional or non-compliant behaviour.

According to the literature, the developmental stages of children with CVI will typically be achieved later in life, as these children experience the world in diverse ways[1,10]. Typically, the following interfering effects can be observed: a) altered gross/fine motor functions, b) deviations in the development of spatial concepts, c) reduced attention and memory, d) underestimation of cognitive abilities, e) uncontextualized behaviour and inappropriate communication skills due to inadequate facial expression interpretation leading to inappropriate social responsiveness (anxiety vs. lack of distance), f) difficulties in achieving specific academic learning processes (reading, writing and calculating), h) bonding disorders and i) disorganisation of sleep-wave patterns. Thus, the clinical profile of CVI is widely heterogenous[7,30].

It must be kept in mind that cerebral visual impairment is a diagnostic umbrella term, which does not indicate which visual functions or capacities are affected and which are not[9]. This is important as it leads to a new understanding of a disease disorder for professionals and must result in specific therapeutic responses in terms of developing appropriate educational materials and shaping appropriate environmental conditions.

4. What kinds of symptoms can be observed in cases of visual subsystem deterioration and inability to execute functions?

4.1 Damage to the posterior visual system

Damage to the **ventral stream** are related to a) impaired orientation within the surrounding environment (topographic agnosia), b) difficulties recognizing peoples' faces as well as difficulties recognizing the language of facial expressions, c) difficulties recognizing animals, d) difficulties recognizing combined objects (i.e. words or numbers), e) difficulties matching visual input with the image of letters and words, f) difficulties with letters that are reversed pairs, such as 'd' and 'b' or 'q' and 'p' (form constancy) g) and the inability to read (numbers or letters) what has one personally written. Reading comprehension might be decreased, because these children tend to think in words rather than in pictures. They have difficulties visualizing and remembering their descriptive text. Some students may struggle with the visuospatial aspects of math; in that respect, that they have no inner notion of numbers[35] (Table 1).

Table 1: Symptoms, dysfunctions and adaptations exhibited by children with ventral stream dysfunction.

Ventral Stream Dysfunction

Symptoms	Dysfunction related to	Adaptation/Behavior
Impaired orientation Difficulty route finding	special recognition	children become easily lost
difficulty recognizing faces	face recognition	look for alternative identifiers
difficulty recognizing facial expression	face expression recognition	autistic behavior/ tantrum temper social isolation, anxiety
difficulty recognizing shapes, figures objects	shape and figure recognition	tactile/smell recognition
difficulty "seeing" the whole picture	"brain accommodation"	short distant viewing
difficulty focusing small items in a whole picture	"brain accommodation"	short distant viewing
difficulty recognizing letters, numbers, words	letter, number recognition	reading and reading comprehension difficulty, avoiding of reading

Damage to the **dorsal stream** are related to a) an impairment in visual searching, which means affected persons cannot simultaneously process multiple elements of a visual scene. Children have difficulties locating the next line of print on a page. Children have difficulties processing one or two visual elements at a time when there is to much print information on a page (so called visual crowding). A child with damage in this area sees as does a baby, who can only concentrate on one thing at a time. This is an apraxia of gaze. In this case, the child is not able to accurately move the head and the eyes to a new location, i.e. it functions not properly and synchronously[36,37]. Children may have a decreased ability to simultaneously process information, while the distracting input can be from auditory, tactile, olfactory, kinaesthetic, mental or physical discomfort. Children may suffer from impaired visual guidance of movement and their visuo-motoric difficulties, which is reflected by clumsiness and/or writing and drawing difficulties. They can show impaired following and tracking of moving objects (apraxia of gaze). This is related to an inability to easily move the eyes from one item to another. Often, children present with decreased spatial awareness (Table 2).

Table 2: Symptoms, dysfunctions and adaptations exhibited by children with dorsal stream dysfunction

Dorsal Stream Dysfunction		
Symptoms	**Dysfunction related to**	**Adaptation/Behaviour**
difficulty finding toys, clothes on a pile difficulty finding objects in clutter	positions	clumsiness
difficulty finding friends/relatives in crowd	visual search	anxiety, insecurity
difficulty jumping/diving in a pool	lengths and directions	anxiety, insecurity
cannot see all the information on a TV screen	decreased pixel processing	prefers slow presentation close to the TV screen
difficulty seeing and listening at the same time	simultaneous perception	temper tantrums, anxiety
items in distance cannot be seen	visual search	decreased risk assessment
impaired reading of crowded text, letters, page	crowding	increase letter/number size and spacing within letters/numbers and well-spaced text line layout
impaired hand writing	guidance of movement	use Laptops

A specific visuo-motoric difficulty in children with dorsal stream dysfunction is difficulty writing horizontally. Lined paper will support this. Additionally, they have difficulties grasping a pen in the correct position in their hands. Children with central visual crowding experience a blur of letters/numbers, or the letters/numbers appear to dance. Children have difficulties seeing all letters in a word or large numbers when there is not adequate spacing between them. These children benefit from limiting the amount of visual information by spacing text or pictures more widely. Some have difficulty increasing and/or decreasing the field of view. Further on, organisation of the workspace and consistent placement of items can helpful.

5. What are the somatic and psychological functions involved and triggered by the different subsystems?

5.1 Visual attention

For a person to see properly, many more of the abovementioned visual processing pathways are needed. Visual attention is needed to register what we see. Visual attention is both conscious and subconscious and is related to visual processing. Without giving attention to the visual system, we do not see consciously. The amount we can see at any moment is limited by the amount that can be processed by the brain at one time and how fast it will be processed. We miss things that move too fast to be seen. Impairment of the conscious attention leads to an apparent lack of focus and concentration. Thus, paying attention to the visual sensory modality is a prerequisite of seeing consciously.

Diverse types of attentional processes turn sensory processing into conscious perception. Our conscious visual attention, which we use to recognize faces, words and numbers, for example, is slower than our subconscious system, because it processes sequentially, focusing on one thing after another. The act of paying conscious attention is regulated via the prefrontal lobes of our brain, which change the amount of attention consciously. Impairment of conscious attention leads to a lack of focus and concentration and errors being made in relation to recognition.

Subconscious visual attention prevents us from bumping into items through fast parallel (simultaneous) processing of the relevant surrounding information. The thalami inside the brainstem help in dealing with the subconscious attentional processes further on resolved by the parietal lobe, which encompasses three automatic, subconscious functional systems: a) the posterior parietal lobes provide the frontal lobe with an unconsciousness framework of the item's location to which attention can be given and choices can be made, b) the posterior parietal lobes provide an internalized 3-dimensional (visuo-spatial) map of the surroundings to visually guided movements and c) the posterior parietal lobes provide an interlink with the memory systems of the hippocampus related to spatial awareness and memory. Impairment of subconscious attention leads to the phenomenon of colliding with obstacles, tripping, being clumsy and an inability to find pieces in cluttered scenes. In the case of damage to both

posterior parietal lobes, one can only see one thing a time, so that only large things can be presented. This is called 'Balint syndrome', which is a complete simultaneous agnosia[27,38].

In summary, damage to any part of the brain related to visual attention can interfere with subconscious or conscious visual attention, depending on which parts of attention and visual brain systems have been damaged. Impairment of subconscious attention may lead to clumsiness or the inability to find people or objects in a patterned or cluttered scene, while impairment of conscious attention leads to a lack of focus and concentration.

5.2 Attention and the executive system

Visual functions cannot present the visual world to us unassisted; they need to be controlled and guided. Both the outside and the inside worlds make use of attention to take control of the brain's systems. By controlling attention, the executive system allows for the selection, at will, of information from the outside world (exogeneous) as well as the inside world (endogenous) of the mind. The executive system controls the sensory systems that provide information from the outside world and the emotional, visceral and cognitive systems forming the content of inside world to achieve or satisfy our basic needs. It additionally rules the motor system, allowing interaction with the outside world. Attention can thus be thought of as a door through which different databases of information—of which vision is one—can be entered. Exogeneous and endogenous attention need to be balanced and controlled. This control is exercised by the executive system inside the frontal lobe[39].

5.3 Visual selective attention processes

In most situations, the incoming information in the visual field is too much to take in at once. The amount of information needs to be reduced or adapted. This is regulated by the visual selective attentional functions. While the eyes can only be directed to one point in space per saccade, visual selective attention can cover an area as large as the whole visual field. This means the size of the attentional area may vary from very small to very large. The ability of the brain to cover this range can be compared to an automatic zoom lens. The left and right temporal areas are connected to the thalami, and it has been suggested that these areas are involved in decreasing and increasing the size of attended visual area (like a zoom lens) to be processed further on to the visual pathways[40]. While the left temporal lobe is involved in decreasing the size by zooming in to cover local visual attention, the right temporal lobe affects global visual attention by increasing the size (zooming-out). Thus, changing the size of the field of view is performed in the thalami, preceding visual processing in the dorsal and ventral streams. These visual selective attention processes determine what is processed to the ventral and dorsal streams. The literature discusses the 'what', 'where' and 'how' pathways, indicating the different higher functional visual features involved. Clinical practice suggests that both functions, decreasing and increasing the size of the field of view, can be disturbed. Children who have a disorder that creates a large field of view (increasing the size by zooming out) have trouble seeing the relationship between details; they lack the overview and often fail to see the ´larger picture`. These children can be unable to identify incomplete visual images as a whole object, pattern or configuration. They would fail in the *Gestalt Closure* test. Simultanagnosia in children can be thus explained as a result of a disturbance of visual attentional functions, i.e. the right temporo-parietal lobe might be affected related to a lack of global selective attentional function.

By contrast, other children might have problems decreasing the size of the selected area and have difficulty zooming in on details. These children have difficulty finding information in crowded visual scenes and reading small text as well ass comprehension problems due to increasing fatigue. They might have trouble accessing multiple details at once, which makes it difficult for them to see details in a crowded visual field, resulting in the experience of blurring of items (letters, numbers etc.) or letters and numbers appearing to dance. This can be evaluated by the 'LEA Test' by Lea Hyvärinen[41]. This crowding effect can be related to a reduced local attentional dysfunction and a reduced *visual brain acuity*. Visual acuity might be measured in the normal range, and very often these children are not recognized by ophthalmologists and considered to be visually normal.

This impairment of subconscious local selective attention leads to colliding with obstacles, being clumsy and having difficulties finding people or objects in crowded or cluttered scenes. Damage to the parietal lobes (dorsal stream) leads to a variety of patterns of visual loss depending on where the damage is located. This so-called 'Balint-Syndrome' is a simultaneous agnosia and is related to damage to both parietal lobes. Historically this has been observed in soldiers with shrapnel injuries to the brain[35,38]. Other observations were that these soldiers were unable to guide movements of the arms, legs and body, which can be described as optical ataxia.

In younger children, cognitive development such as reading/writing achievement may be disturbed if there is a simultaneous agnosia[42]. Additionally, an inability to move the eyes to look at various parts of the surrounding scene is described, which can be identified by saccade measurements. Patients may suffer from problems with visual guidance of movement and/or visual size variation (visual selective attention). Patients do benefit from increasing the size of the print and decreasing the amount of less information on the page as well as increasing the spacing between lines and words.

5.4 Visual imagery and mental visual manipulation

Mental imagery and mental manipulations of the visual information is regulated by the working memory in the prefrontal cortex, not in the visual cortex. Non-verbal working memory allows children to represent and visualize objects and places in the outside world, even when the objects are not present.

The ability to have object permanence means that representations of objects continue to exist even when they are not seen. In order to look for something, the mind needs one or more active representations or visual images to which visual stimuli can be matched. Representation of objects, features of objects and faces in the ventral stream are used to provide and match the image of the target. The prefrontal cortex allows the mental imagery and mental manipulations of visual information which is stored in the ventral and dorsal stream. Thus, only the contextual information is taken from the ventral and dorsal pathways of the visual cortex. The ability to call on visual images and to keep them active in the visual working memory is considered an executive function which takes place in the prefrontal cortex. The executive system in the prefrontal lobe is thus involved in visual problem solving[39,43].

Visual closure is a feature used to mentally fill in gaps in incomplete visual objects or pictures. This is either related to the ability to build mental visual images in the prefrontal cortex and/or to increase the size attending to the whole stimulus area in the temporal lobe (see global visual selective attention). Thus, visual closure is an ability which requires two higher visual functions: First, the child should be able to address the entire stimulus area (global visual selective attention). Second, it should be able to engage in visual imagery to actively fill in large gaps in visual objects or pictures. An impairment in the ability to form mental images leads to difficulty giving meaning to the world. In the case of a CVI, the outside visual world may interfere with the ability to generate mental images. In this context, distinct functions must be considered: First, information has to be actively recruited and retained in the ventral stream. In a subsequent step, the image can be sustained in the mind and subjected to further mental manipulation. The clinical consequences of a problem of visual imagery are a) being unable to create images or manipulate images (i.e. difficulty drawing of an image from the memory), b) difficulty finding missing objects (visual closure), c) and inability to retrace a route by remembering landmarks. These difficulties result from general working memory disorders that affect visual task performance. Thus, executive functions control either general or visual attentional processes, which determine what is being processed in the dorsal and ventral stream pathways, and active mental imagery and manipulations of visual images, which play a role in providing a contextual background against which visual input is interpretated.

6. What are the assessment procedures for the abovementioned symptoms and diagnoses?

Often, clinical difficulties result from complex interactions among visual, attentional, executive or other dysfunctions. Thus, the provision of appropriate therapeutic responses requires the assessment of visual, attentional, executive, motor, cognitive and other sensory functions. Assessment of visual dysfunctions cannot be reduced to the traditional components associated with ocular visual impairment but must be extended to include higher levels of integration of attentional, perceptual and visuo-spatial functions[5,44,45]. Neurophysiological tests can be used to determine the quality of lower- and higher-order visual functions. These functions are the essential building blocks provided by processes of the brain that give rise to and determine the quality of visual experience and behaviour.

Assessment of functional vision is not conducted in standard ophthalmic practices. It extends beyond the issues related to visual acuity and the visual field. Functional vision describes how an individual functions using vision and involves evaluation of that person's vision skills and its application to the performance of visual tasks (i.e. reading, writing, drawing, painting, moving, communicating etc.)[46]. Visual functions refer to and describe how the ocular and cerebral visual systems function together, with measures taken at the eye and brain levels. In many children poor performance on visual tasks results from a complex interaction of executive, visual and other functions, which take place inside the thalami and the occipital lobes and

further on in the ventral and dorsal streams. Damage to the occipital/temporal/parietal lobes and the thalami decrease the probability of clinical symptom improvement significantly.

Specifically, professionals should be interested in capturing the following information, and often family members are the best source of information about their children's behaviour[46,47].

a) General capacity to maintain visual attention; b) visual selective attentional function, which concerns global and local attention and takes place inside the thalami and the temporo-parietal lobes; c) visual perceptual function, which concerns visual consciousness and identification of objects, faces and images; d) visual memory function (spatial orientation and how fast objects are moving in which direction), which takes place in the ventral steam; e) visual working memory function, which is important for visual imagery, which means being able to generate and manipulate a mental visual image, and takes place in the prefrontal cortex; f) guidance of movement (visual-motor function), which takes place in the dorsal stream; g) visual emotional function (face and gesture interpretation), which takes place in the ventral stream and h) visual processing speed (being able to and perceive visual stimuli quickly) in all areas.

Non-verbal messages may be misinterpreted as a result of the inability to see or interpretate gestures or body language or the inability to access information due to associated auditory processing difficulties. This can be an inability to discriminate one face from another and associate the face with a name (prosopagnosia) or a difficulty interpreting facial expressions and body language. Difficulty negotiating crowded areas as a result of difficulty with spatial orientation or not being able to visually perceive individual people or objects crowded together may lead to an impression of anxious or fearful behaviour.

6.1 CVI-related questions for use in early clinical check-ups

The following checklist (questionnaire) should be considered as an instrument for detecting possible visual abnormalities:

Does your child have difficulty walking downstairs? Does your child have difficulty seeing things that are moving quickly, such as small animals, or does your child react in an irritated or fearful way to fast-moving animals? Does your child have difficulty seeing objects that are pointed out in the distance? Does your child have difficulty locating an item of clothing in a pile of clothes? Does your child find copying words/numbers or drawings time consuming and difficult? Is there a lack of awareness that their reactions in social situations are atypical? Is there a lack of awareness of the reactions of others to what they say or do? Is there a lack of awareness of how others perceive them? Is there a lack of awareness that they may perceive the world differently to others? Does your child have difficulties understanding the meaning behind words and phrases (pragmatics)? Does your child give the appearance of indifference and lack of interest? Does your child engage in unconstructive behaviours when faced with difficult situations? Is there possibly a overreliance on others for support and direction? Does your child have difficulty with trust? Is your child involved in bullying, either as a victim or as a culprit?

7. What are appropriate causal general and specific medical interventions?

7.1 Academic interventions

There is little information about CVI in the psychological and medical literature. The following types of interventions are of great importance. First, early identification of CVI is of great relevance as the sensory building of cerebral networks begins shortly after birth. So, the focus should be on integrated sensory visual information. The therapeutic focus should include the contribution of the family in this context. In the case of a child misdiagnosed as a result of other conditions, inappropriate intervention strategies may be used and negatively affect the child's academic and general progress. Possible academic intervention skills in school are basically orientated toward literacy, writing, and math skills: i.e. increased letter size, increased line spacing, screen magnification and phonetic support (hearing books). For listening comprehension, it would be beneficial to make textbooks available in audible form. It can be beneficial for children to dictate answers to a worksheet or a test. A computer could be used for all writing tasks. For mathematics, visual support can be provided by using pictograms or graphic presentation, using graph or square paper for long calculations, occluding irrelevant information on the paper and providing use of a video magnifier.

7.2 Psycho-social interventions

Communication skills are essential in children with CVI. They can be supported by giving additional verbal cues. Voice recognition can assist with visual searching. In some cases, it might be necessary to wear brightly coloured items, which pop out from the background. It must be ensured that school physical education staff is aware of the difficulty children with CVI encounter playing team games with many people moving. For this reason,

individual sporting activities involving fewer people (e.g. athletics and swimming) should be chosen.

7.3 Specific medical interventions

Depending on the localization of the damage, various kinds of interventions (for example, by application of a prism or coloured glass or by medication) may apply, e.g. inside the ventral stream, special areas are responsible for colour processing. If, for any reason, processing of the different wavelengths might be affected, this can be addressed by using coloured glasses that potentially absorb the specific wavelength which interferes with regular processing[48]. Prismatic correction might correct the retinal structure on the way through the thalamus to the occipital lobe to correct any interferences during processing, while a medical stimulant might help in regulating thalamus gating[49]. All these interventions refer to a stimulation of different fibres on their way to the thalami, thus possibly opening the thalami channels and giving access to a controlled binocular saccadic movement. In our experience, neurophysiological training can be applied as a therapeutic tool to gain access to mental imagery constructions and the prefrontal support of visual attention.

7.4 CVI vs. congenital blindness

In case of congenital blindness reorganization of the brain is performed through activation of the somatosensoric cortex by using the tactile information through Braille lettering. Additionally, activation of parts of the occipital visual brain centers for processing the tactile information of the Braille points[50]. This is different from what should be achieved compared to CVI treatment.

8. Further directions in the medical treatment of CVI

Manifestations of CVI in children are represented as a medical spectrum diagnosis, ranging from profound to mild and leading to differential management interventions in the medical, psychological and academic environments. Unfortunately, in all these environments, knowledge of this medical diagnosis is very rare as professionals are often unaware of the features and needs of these children. Performance in visual tasks can diminish in the context of fatigue, distraction and stress, as there are no functional reserves. We have to be very careful not to misinterpret behavioural symptoms, as this can lead to criticism from the caregiver's side and lower self-esteem for the child, who cannot understand what is 'wrong'.

The pathology of CVI in the visual brain leads to distinct features in children compared with adults. We cannot apply adult modes of interpretation of the features of perceptual and cognitive visual dysfunctions in children as the exploration must consider the developing character of the brain in childhood. As this is an early onset of damage, it affects a range of interacting and developing brain functions. Considering the neuroplasticity of the developing brain, CVI may lead to variable degrees of recovery. Additionally, in children, compensatory behavioural adaptational symptoms mask the underlying deficit. Children have not consciously perceived another type of vision and believe their vison to be normal. They have no means of comparing themselves with their healthy peer group. Their observed behaviours can be related to their visual disorders with informational support from parents and other caregivers by taking a structured CVI-related history. In general, the course of check-up examinations in children must consider the contribution of **seeing** to communication (person to person and group), orientation and mobility in space, life and every day handling, tasks which require sustained near vision (i.e., painting, reading, writing etc.), the so-called cultural abilities. In this context, we must consider three situations: 1) What is the quality of the visual picture the child sees? 2) How does the child process information using higher visual functions? 3) How do visual pattern deviations influence the development of the child?

Further research is necessary to embed different developing visual/auditory functional systems in children under the diagnostical and therapeutic umbrella.

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Conflict of Interests

The authors declare that they have no conflict of interest.

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