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Impairment of vision in children due to damage to the brain: a practical approach

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Abstract

Aim: **To describe the visual problems which children with brain damage may experience and to describe approaches to these, applicable to everyday life.**
Methods: **A literature-based essay is presented using relevant articles and practical experience.**
Results: **Damage to the brain can affect both visual input and visual processing. Degradation of visual input results in reduced visual acuity, reduced contrast sensitivity and restricted visual fields. The dorsal stream passes between the occipital cortex and the posterior parietal lobes, which serve the ability to extract visual data from a complex scene, visual attention and the visual guidance of movement. The ventral stream passes between the occipital lobes and the temporal lobes and is responsible for recognition of what is being looked at and orientation. Any of these visual functions can be impaired to any degree and in any combination. Structured history-taking and clinical examination is required to fully characterise the visual problems in each affected child in order to devise a tailored management strategy matched to the child's needs.**
Conclusion: **Structured history-taking and clinical examination in children with visual impairment secondary to brain damage can allow management to be customised to the individual requirements of that child.**

Key words: Cerebral visual impairment, Cortical visual impairment, Dorsal stream, Ventral stream

Introduction

Our vision is served by an input system and a processing system. The input system connects the eyes to the occipital lobes and is well understood by the ophthalmic professions.[1] However, the processing system is less well appreciated despite patients presenting with a wide range of complex disorders of vision due to impaired visual processing, which is the focus of this paper.

Damage to the visual brain can impair visual acuity and contrast sensitivity, and may restrict visual fields,[2,3] while damage to the higher visual processing centres causes perceptual and cognitive visual impairment. Such damage may affect the outer brain tissue (grey matter or cortex) or the inner brain tissue (white matter), and lead to visual dysfunction, which varies in nature and degree. In children this can cause multiple problems including impaired recognition of people, shape and objects, orientation problems, difficulty handling complex visual scenes and inaccurate visual guidance of the limbs.[4–8] The visual system may be the only part affected, or there may be associated damage to other brain structures, resulting in cerebral palsy and/or other developmental problems. The term cortical visual impairment, used to describe visual dysfunction from cortex damage, is rare in isolation. The term cerebral visual impairment (CVI) is preferred to encompass damage to the grey and white matter of the brain.

CVI, which is now the commonest cause of visual impairment in children in developed countries, is increasing in prevalence due to improved perinatal care and survival of young children with profound neurological disease.[3] Many causes of brain damage in early childhood can produce CVI. The most common are periventricular leucomalacia[9] (damage to the white matter surrounding the lateral ventricles, seen in preterm infants, who may also manifest impaired cortical development[10]), hypoxia and ischaemia, hydrocephalus,[11] meningitis, encephalitis, traumatic brain injury, metabolic disease and the secondary effects of drugs and radiation.[12–14]

The visual system

The primary visual pathway begins with light entering the eye and stimulating the retina. The visual information passes along the optic nerves, chiasm and optic tracts to the lateral geniculate nuclei, then via the optic radiations, to the occipital lobes.

Higher visual processing takes place in adjacent brain areas. Two pathways are central to this process: the dorsal stream and ventral stream. The dorsal stream runs

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between the occipital lobes and the posterior parietal lobes, which subconsciously appraise the whole visual scene along with all other sensory inputs, providing the facility for the frontal lobes to choose the components to pay attention to. This area computes the location of components of the visual scene and thereby facilitates visual guidance of movement by passing the coordinates of three-dimensional visual space to both the motor cortex to plan and bring about movement of the body, and the frontal eye fields to generate rapid, accurate head and eye movements to chosen targets in the visual scene. A part of the scene is chosen, for example an apple, and the information is passed to the frontal lobes, which instruct the head and eyes to look at it. The apple's coordinates are passed to the motor cortex, which initiates hand movement and accurate reach with pre-adjustment of finger position, to grasp the apple. (The 'picture' of the apple is inside the brain. The miracle of vision is that this becomes coincident with reality, so the apple is lifted by successfully shaping and moving the hand to emulate the 'picture' within the mind).[15]

The ventral stream connects the occipital lobes to the temporal lobes, which contain the brain's 'visual library'. Information transmitted here allows recognition and visual memory of what is being looked at. Recognition of faces involves image data passing along the ventral stream into the temporal lobes (commonly the right) where it is compared with data concerning all known faces stored in the fusiform gyrus. If there is a match, the face is recognised. Recognition of shape and form and the ability to recognise and follow routes are likewise temporal lobe functions.

Damage can affect any part of this overall visual system in any combination and degree, giving rise to a wide range of patterns of visual dysfunction.[13,16,17]

The features of visual dysfunction

Visual acuity

Visual acuity (VA), or clarity of vision, is easily recordable; however, this does not necessarily reflect functional vision. It is a measure of the resolution of the visual system and is commonly (but not always) reduced in CVI, in the presence of normal pupil responses and normal fundi. Binocular VA is measured to determine visual ability; however, the functional visual level may be more dependent on the level of concentration and attention. To discover the size of text that can be seen and understood when the child is tired, functional VA needs to be investigated. Visual crowding is well recognised in amblyopia. A similar, and often more severe, variant of the same phenomenon can be seen in children with dorsal stream dysfunction.

Contrast and colour vision

Colour vision is often normal in CVI, although accurate testing can be problematic due to other disabilities. Contrast sensitivity, on the other hand, can be significantly reduced in children with CVI.

The visual field

Many visual field defects can occur. Unilateral damage to the retrochiasmal visual pathways results in field loss on the opposite side, either a complete or incomplete homonymous hemianopia. Bilateral damage to the posterior visual pathways, above the lateral ventricles, causes lower visual field loss. These patterns of visual field loss commonly result from injury in the perinatal period, and a degree of spontaneous recovery can take place. Insults to the visual pathways *in utero* may lead to diffuse impairment of visual function in the affected area.

A false impression of visual field impairment can be given by dorsal stream dysfunction. Unilateral posterior parietal damage, particularly on the right, gives rise to lack of attention on the opposite side, emulating hemianopia. Bilateral posterior parietal pathology causes an inability to see multiple targets simultaneously (giving the impression of visual field constriction) and/or a difficulty in estimating depth for guidance of motion, causing clumsiness, which may be interpreted as lower visual field loss.

Perception of movement

The perception of movement centres are found in the middle temporal lobes in the region of the occipito-parietal temporal junction. Bilateral damage can cause impaired or absent perception of movement. Only the static visual world is appreciated in this situation. In contrast, individuals in whom this area is intact despite severe damage to the occipital lobes may exhibit movement perception as the only visual function, because function persists in this area in addition to the areas responsible for reflex visual perception, the superior colliculi and the pulvinar.[15]

Recent research has shown that damage to the peri-ventricular white matter in the parieto-occipital region causes difficulties with perception of motion required to recognise biological movement.[18]

Impaired accommodation

Many infants and young children are hypermetropic; blurred images stimulate accommodation resulting in clear vision. Incomplete accommodation is common in CVI and the persistent hypermetropia and consequent blurring of the retinal image is potentially amblyogenic. Refractive correction of even small degrees of hypermetropia can lead to significant improvement in vision. The additional magnification from hypermetropic lenses may also compensate for reduced acuity and visual crowding, and it is worth eliciting whether this is the case. Lack of accommodation is seen in over 50% of children with cerebral palsy, many of whom benefit from near spectacle correction. Thus consideration should be given to screening all such children, by dynamic retinoscopy, for lack of accommodation.[19]

Impaired eye movements

Common eye movement problems in CVI include strabismus, nystagmus, unstable fixation, inaccurate fast eye movements (dysmetric saccades), deficient smooth pursuit movements and paroxysmal deviations. Strabismus can cause amblyopia, with impaired binocular

Table 1. The features of dorsal stream dysfunction

Feature	Problems
Impaired ability to handle complex visual scenes	Finding a toy in a toy box
	Finding an object on a patterned background
	Finding an item of clothing in a pile of clothes
	Finding food on a plate
	Seeing a distant object
	Reading
	Identifying someone in a group
	Tendency to get lost in crowded locations
Impaired visually guided movement	Floor boundaries
	Uneven surfaces
	Steps
	Kerbs
	Inaccurate visually guided reach
Impaired attention	Performing more than one visual task at a time
	Marked frustration at being distracted

vision and stereopsis. CVI may also affect the eyes' ability to move quickly from one area of interest to another (psychic paralysis of gaze), impairing the tracking of moving objects, particularly if fast. Difficulties with visually guided eye movements may be compensated for by head movement in capable children. Reading may be impaired because of deficient small movements of the eye required to read one word after another.

The implications of dorsal stream dysfunction (Table 1)

The ability to handle complex visual data increases with age. If this is dysfunctional the child may demonstrate difficulty in finding an object of interest if the visual scene is too complex, for example a toy in a toy box or on a patterned bedspread.[4] A similar problem arises with food mixed together or presented on a patterned plate. Seeing objects in the distance is difficult, as the further away things are, the more information there is in the visual scene. Busy environments such as supermarkets, shopping centres or swimming pools present a complex visual scene. Children may react in different ways, becoming either frightened or disruptive. Reading may be affected, particularly when print size diminishes with each school year. The progressive crowding of the text increases the complexity of the visual scene.

Inaccurate movement through three-dimensional space may occur. Mobility problems result from an inability to differentiate a floor boundary (for example between carpet and linoleum) from a step. The child is reluctant to cross it before careful exploration. Black and white tiled floors can be frightening and large patterns on carpet difficult to cross. Stairs and kerbs are problematic as the foot is lifted to the wrong height, too early or too late. Going down stairs is particularly difficult because of difficulty in judging depth. Inaccurate reach and grasp may also manifest. Such impairment of visually guided movement is called optic ataxia.

Attention is often impaired. This becomes evident when the child has to perform more than one task at once, such as talking and walking. This can cause marked frustration at being distracted.

Table 2. The features of ventral stream dysfunction

Feature	Problems
Impaired recognition	Recognising people in person or in photographs
	Interpreting facial expression
	Recognising animals
	Recognising shapes and objects
Impaired orientation	Getting lost in known locations and new environments

The implications of ventral stream dysfunction (Table 2)

Recognition problems commonly result in a difficulty identifying faces.[4] The child may be unable to recognise even close family members or people seen out of context. This may lead to identification of strangers as being known. Identification of people in photographs is challenging, as is seeing, interpreting and understanding facial expressions. Similar difficulties in identifying animals, objects, shape and form occur.

Orientation problems are common, affecting route-finding. The child may get lost and require help, even in well-known locations. The problem is more marked in new environments, such as shops or hotels, resulting in the child becoming frightened, withdrawn or disruptive.

The diagnosis of CVI

CVI manifests in many ways. Any of the cerebral visual functions may be affected, in any combination and to any degree. Problems experienced by individuals vary widely, encompassing any of the problems previously described. In children with marked CVI or other neurological problems the diagnosis may be obvious. In children who are otherwise apparently normal, the symptoms and signs may be more subtle and the diagnosis must be considered and explored. Eliciting the problem requires detailed history-taking to encourage parents to report the problems. Structured questioning, targeting the recognised manifestation of CVI, is required.

The management of CVI (Table 3)

The aim of management is to identify the problems each individual child has and to find practical solutions. Parents often adopt novel ways to aid day-to-day living. Much of management understanding comes from their reported experiences. Conveying information to families is central to developing suitable coping strategies and to adapting to future problems.[20]

Reduced functional **visual acuity** is helped by increasing the size and/or proximity of text and images. Print can be enlarged, double-spaced and presented in small sections. Visual acuity may improve by limiting distractions and avoiding tiredness. The thickness of the felt-tip pen or pencil used should match the acuity. (The thickness of the lines of Cardiff card images provides a guide.)

Children with **poor visual acuities** and **contrast**

Table 3. Management strategies for children with CVI

Problem	Solution
Reduced functional visual acuity	Enlarge text Double-space text Present text in small sections Reduce distractions Limit tiredness
Colour vision and contrast sensitivity impairment	Bright and clear educational material and toys Distinct colour boundaries Good contrast
Hemianopia	Tracing of text with a finger or ruler Turning text vertically or obliquely Appropriate seat position in classroom Turning of head to check the hemianopic side Careful guidance around new environments Training in crossing roads Turn plate to eat food
Lower visual field defect	As with hemianopia Regularly looking down to check the ground ahead Tactile guide to ground height
Impaired tracking	Movement of the head Enlarging text Double-spacing text Tracing of text with a finger or ruler
Impaired movement perception	Television programmes with limited movement Educational material with limited movement Careful training or guidance in crossing roads
Finding a toy in a toy box; finding an item of clothing in a pile or wardrobe	Separate storage of favourite items Organised storage systems Always store in same location Avoid clutter Colour coding and labels
Finding an object on a patterned background	Use plain carpets, bedspreads and decoration
Finding food on a plate	Avoid patterned plates Avoid sauces/gravy Separate food portions
Seeing a distant object	Use zoom on video/digital camera to view
Reading	Enlarge text Double-space text Masking surrounding text Computer programs to present information
Identifying someone in a group	Wear an obvious identifier Always stand in the same location Wave Speak
Tendency to get lost	Training in seeking and identifying landmarks Visit new locations at quiet times
Problems with floor boundaries, steps, kerbs and uneven surfaces	Avoid patterned floor surfaces Banister Mark edge of stairs Good lighting Tactile guides to gauge the height of the ground Approach obstacles with 'Look, Slow, Check, Go'
Inaccurate visually guided reach	Activities to improve coordination Reaching beyond an object to gather it Activities to improve coordination Occupational therapy
Difficulty 'seeing' when talking at the same time	Limit conversation when walking Identify obstacles by tactile stimulation
Frustration at being distracted	Limit distraction Reduce background clutter Reduce background activity Quiet table at school
Difficulty recognising people and photographs	Introductions Training in identifying voices Consistent identifiers worn Training to recognise identifiers
Difficulty recognising shapes and objects	Training to identify and recognise identifiers Training in tactile recognition
Difficulty reading facial expressions	Training in recognising facial expressions Expression of mood by tone of voice Explanation of mood in words
Getting lost in known locations	Training in orientation Encouragement in leading Incorporate landmarks in mnemonics/poems
Difficulty in new environments	Training in orientation Encouragement in exploration: visit at a quiet time; hide and seek; treasure hunts
Visual fatigue; prolonged visual processing	Minimise clutter Reduce distractions Reduce detail and complexity Well-earned breaks
Social problems	School support Identify problems and solutions Encourage child to overcome them Well-known peer group Find activities child enjoys and can excel in

sensitivity difficulties require toys and educational material to be bright and clear. Distinct colour boundaries and good contrast are essential.

Visual field defects affect reading, writing, communication, mobility and eating. **Hemianopia** restricts reading. Right hemianopia causes each new word to jump into view and may not be anticipated when looking straight ahead. In left hemianopia the text progressively disappears, so locating the start of the line below is tricky. Using a finger to trace the words and reading text vertically or obliquely so the hemianopia covers what has just been read is helpful (reading vertically downward for left hemianopia and vertically upward for left hemianopia). Writing is aided by a ruler guide to progress along a line.

Hemianopia can cause significant communication problems. The child is unaware of people sitting or approaching from the affected side. Position in the classroom needs consideration (children with left hemianopia need to sit with the teacher slightly to their right and vice versa).

Mobility problems in hemianopia include the child bumping into obstacles on the affected side. They need to be taught to turn their head to check the hemianopic side. Careful guidance around a new environment may be required. Training to fully turn the head before crossing roads is necessary for safety.

Hemianopia may cause a child to leave food on one half of the plate. The child must learn to turn the plate. Exploration within the field defect can be encouraged by placing favourite foods on the hemianopic side.

Visual field extinction is identified when a child sees a moving finger on each side but not on one side when the other is simultaneously moved in equal degree in the mirror image position. We have seen two children with this condition who panicked in long straight corridors. We hypothesised that it was the symmetry of the corridors that caused the problem (as one side could seem to disappear). The problem was eliminated by decorating one side of the corridors at school with posters.

Lower visual field defects cause analogous problems to hemianopia and comparable coping strategies can be employed. In particular mobility is affected due to an inability to see the ground ahead. The child must be taught to look down regularly. A tactile guide can be useful to feel the ground ahead. A tilted workstation enhances access to information at the bottom of the page.

Visual inattention on one side resembles visual field impairment but no field loss can be identified. Features include writing on only one side of the page, a tendency to bump into doorframes on one side and a tendency to veer consistently to one side. In some cases drawing may be impaired on one side of the picture only. Inattention of the left visual scene tends to be more severe, and thus more commonly manifests. Compensatory strategies include presenting text off-centre to the unaffected side, ensuring the teacher is on the unaffected side, and placing a visual distracter (such as brightly coloured tape) at eye level on the doorframe on the affected side. Drawing can be carried out one side at a time with the affected side being drawn upside down, placing it in the intact functioning visual field.

Impaired tracking affects the following of a moving target, compensated for by movement of the head, if the object is moving slowly. Enlarging and double-spacing text and using a finger to trace the words aid resultant reading difficulties.

Impaired movement perception means only slow-moving or static objects are seen. Television programmes with limited movement are preferred, such as the weather report or news. Cartoons with rapid movement can frighten the child. Educational material must be adapted accordingly. Safety can be compromised by an inability to see moving traffic, and extensive training or guidance in crossing roads is essential.

The aim of management of **impaired ability to handle complex visual scenes** is to decrease the amount of incoming visual information. The child may be unable to locate a specific toy in a toy box or an item of clothing in a pile. Clutter avoidance with organised storage systems where everything is clearly laid out helps. Favourite items being stored separately allows the child to locate these independently. Items should be kept in the same location and the child informed if they are moved. Colour-coding and labelling of items assist the child to locate a desired object, facilitating independence. As a patterned background increases visual information, plain carpets, bedspreads and decoration are useful. Food presented on a patterned plate or mixed with sauces or gravy may not be seen. Plain plates with separate food portions may help.

The viewing of distant objects is aided by a zoom lens on a camera to focus on the object of interest. Large blocks of text present too much information and type should be enlarged and double-spaced. Masking of surrounding text with a hand, piece of paper or typoscope, or using computer programs to present small amounts of sequential information, help reading. In class, a transparent pencil bag improves performance. The workstation needs to have limited distraction and the teacher must stand against an uncluttered background during group activities. Time out, in a calm environment, may be needed. Other children in the class need to be educated about reduced vision and how to behave appropriately.

Recognising people, especially within a large group, is helped by the person wearing a consistent identifier, always standing in the same location, and attracting the child's attention by waving or speaking. A buddy system in the playground, and recognition that the affected child cannot identify his or her peers, are essential.

A tendency to get lost, even in well-known locations such as school corridors, is most obvious when these are busy and crowded. Training to seek and identify landmarks to reorientate the child is required. New environments necessitate careful exploration, which is easier if they are visited first at a quiet time, for example closing time at a supermarket, and in advance of going to parties.

Impaired visually guided movement affects the lower and less commonly the upper limbs. An inability to distinguish a floor boundary from a step and/or problems with uneven surfaces result. Patterned floor surfaces should be avoided. Tactile exploration with a foot is compensatory but guides to gauge the height of

the ground ahead, for example a toy pram or bike, are more socially acceptable. An affected child tends to hold onto the clothes of an accompanying adult while pulling down; this provides a tactile guide to the height of the ground ahead. When walking holding hands, parents need to be taught to straighten their arm and hold it slightly back, providing the same guidance with advance notice. Steps, kerbs or obstacles are often approached with caution or apprehension. A banister helps considerably, providing tactile and proprioceptive depth information, as does good lighting and marking the edge of the stairs. A child can tackle an obstacle with the memorised phrase 'Look, Slow, Check, Go'. Inaccurate visually guided reach is overcome by reaching beyond an object to gather it in. If the object being handled is in contact with part of the body this gives guidance for its location so that reach becomes more accurate. In general impaired visually guided movement is helped by activities to improve coordination, such as dancing or martial arts and occupational therapy.

Impaired attention causing difficulty in performing more than one visual task at a time may manifest as an apparent difficulty in seeing when talking at the same time or a marked frustration at being distracted. Conversation should be limited when walking and obstacles ahead identified by tactile stimulation. Distraction, background clutter and background activity should be limited to allow concentration on one objective, for example a quiet table where the child can work alone. At school, recognition of how much the child is able to handle at any one time facilitates the development and delivery of the curriculum.

Impaired recognition of people is overcome if they always introduce themselves to the child and wear consistent identifiers. A class teacher may be distinguished from other teachers in the school by wearing the same item of clothing or jewellery. The child can be trained to identify people by recognising their voice. Training to identify people in photographs, animals, shapes and objects is required. The child must learn and memorise a specific identifier for each of these.

Training in tactile recognition is valuable. Difficulty reading facial expression is very socially disabling. Training of the meaning portrayed in pictures of classical facial expressions may be useful. Feelings and sentiment should be conveyed to the child by the appropriate tone of voice or explanation in words.

Impaired orientation leads to the child getting lost in known locations, with a heightened problem in unfamiliar environments. Training in orientation allows independence as they grow older. This is achieved by encouraging the child to lead when they are out with a parent and to explore safe areas alone. Games, such as hide and seek and treasure hunts, allow them to gain confidence in orientation. A child may travel a well-known route alone by incorporating landmarks into a mnemonic or poem that is memorised and recited. New locations are best visited at a quiet time to allow exploration with minimal distractions. With familiarity they become more comfortable with background activity.

These children suffer from **visual fatigue** in all aspects of their lives, due to a prolonged visual processing time. In general this is helped by minimising clutter, reducing distractions, detail and complexity and ensuring the child has plenty of well-earned breaks

CVI can cause multiple **social problems** and children are often labelled as being disruptive and noisy. Conversely they may become isolated and appear withdrawn. An understanding of CVI is essential for all those involved in the everyday life of the child. School support often greatly enhances learning and improves behavioural problems. Problems must be identified and solutions found. Time must be invested in implementing these effectively, with constant encouragement. Finding activities the child can enjoy and excel in and having a well-known supportive peer group is invaluable.

Conclusion

CVI is an increasingly recognised problem, encompassing a wide range of problems due to impaired processing of visual information. This may be manifested by impaired visual acuity, colour vision, contrast sensitivity, visual fields, movement perception, tracking, ability to handle complex visual scenes, visually guided movement, visual attention, recognition and orientation. A prolonged visual processing time means visual fatigue occurs. Each child displays a range of problems to varying degrees, but all are associated with social problems. CVI should be considered as a diagnosis when any of these symptoms are apparent. Structured, detailed, focused history-taking may elicit a group of symptoms suggestive of CVI. The specific problems of an individual child should be identified and solutions adopted to overcome these. Improved recognition with better access to information, resources and support is essential for these children and their families and can greatly enhance all aspects of their lives.

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