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Review Article

Visual disorders in children with cerebral palsy: the implications for rehabilitation programs and school work

Gordon N. Dutton [a,c,*], Julie Calvert[b], Deborah Cockburn[d], Hussein Ibrahim[b], Catriona Macintyre-Beon[c]

[a]*Tennent Institute of Ophthalmology, Gartnavel General Hospital, Great Western Road, Glasgow G12 0YN, UK*
[b]*Department of Vision Sciences Glasgow Caledonian University Cowcaddens Road Glasgow G4 0BA, UK*
[c]*The Royal Hospital for Sick Children, Yorkhill Glasgow G3 8SJ, UK*
[d]*Southbank Child Centre 207 Old Rutherglen Road , Gorbals, Glasgow, G5 0RE, UK*

Abstract. Damage to the brain is the most common cause of visual impairment in children in the developed world, and many children with cerebral palsy are affected. The severity varies. Profound visual impairment results from bilateral occipital lobe damage. Visual acuities are frequently impaired but may be within the normal range. Accommodation is commonly impaired and requires appropriate correction. Limitation of the visual fields includes hemianopia due to unilateral damage, and lower visual field impairment due to periventricular white matter pathology. Perceptual visual dysfunction includes impaired visual guidance of movement (optic ataxia), associated with impaired visual search and attention, (due to posterior parietal / dorsal stream dysfunction) and impaired recognition and orientation (due to temporal lobe / ventral stream dysfunction). Impairment of eye movements may also contribute to the clinical picture. Structured evaluation of all aspects of visual function, matched to each child's condition and construction of an optimal management plan, (which can be understood and implemented by everyone looking after and teaching the child), is needed to ensure that no child with cerebral palsy is inappropriately disadvantaged on account of their additional cerebral visual impairment.

Key words: Cerebral visual impairment, cortical visual impairment, vision

1. Introduction

The term 'cerebral visual impairment' (CVI) encompasses visual pathway disorders which impair visual acuities and visual fields, visual disturbance on account of oculomotor incoordination, and visual cognitive and perceptual impairment owing to pathology affecting the visual association cortices and their interconnecting pathways (1). The visual brain is so complex, that disorders during development cause a wide of range of manifestations, which vary in nature and degree. CVI is common in children with cerebral palsy (2), as are refractive error (3) and accommodative dysfunction (4). A large proportion of the brain is devoted to vision, but unlike cerebral palsy in which impaired limb movement is evident (5), cerebral visual dysfunction is less apparent, because CVI is an internal deficit and may go unrecognized unless the resultant visual difficulties are actively sought (6). Apparent developmental disorders may be incorrectly ascribed to disordered mental processing, rather than inability to access and process visual information. Identification, characterization and measurement of visual dysfunction in children with cerebral palsy must be identified, and each element should be measured, and characterized to optimize education and habilitation (7).

This article outlines the clinical evaluation of the visual performance of the child with cerebral palsy, and suggests habilitational strategies for both home and school matched to the nature and degree of each of the visual difficulties identified.

[*]Correspondence: Gordon N. Dutton
Tennent Institute of Ophthalmology, Gartnavel General Hospital, Great Western Road, Glasgow G12 0YN, UK
Tel: 0141 211 2937 Fax: 0141 211 6290
dutton@ledcresc-adsl.demon.co.uk

2. The visual processing system

The process of vision commences 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 (8) where primary processing of the visual image data takes place.

Higher visual processing takes place in adjacent brain areas. Two pathways, the dorsal and ventral stream, are central for this process, (9). The dorsal stream runs between the occipital lobes and the posterior parietal lobes (10), which subconsciously appraise and accord a cognate location for the whole visual scene, along with other sensory modalities (11-13), 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 the surrounding imagery in 3D visual space to both the motor cortex to plan and bring about movement of the body, and to the frontal eye fields to generate rapid, accurate head and eye movements to chosen targets in the visual scene (14,15).

A part of the scene is chosen, for example an apple, 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). The posterior parietal lobes also serve the function of considering all the elements of the visual scene so that attention can be given to any single element, allowing items, which are being sought, to be found.

The ventral stream connects the occipital lobes to the temporal lobes, which contain the brain's 'visual library'. Information transmitted here and serves recognition and visual memory for what is being looked at. For example, recognition of faces involves image data passing along the ventral stream into the temporal lobes (commonly the right) where they are 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 also temporal lobe functions. Damage can affect any part of this overall visual system, giving rise to a wide range of patterns of visual dysfunction (16-18).

3. Visual problems in children with cerebral palsy

Refractive error and impairment of accommodation

Emmetropization is the process by which refractive errors diminish, and normal sight is gained, as a child grows and develops. Impaired emmetropization is common in children with cerebral palsy and this contributes to the high incidence of refractive error requiring spectacle correction. Correction of small amounts of hypermetropia is indicated for children with difficulty of reading, as the spectacles also magnify, and this reduces visual crowding of text (19).

Accommodation is the automatic process of bringing the eyes into focus when they look at a near target, and this is synchronized with convergence of the eyes, which is required to give single vision for near. Accommodation is frequently impaired in children with cerebral palsy. In a comprehensive survey of children with cerebral palsy in Northern Ireland over 50% showed significantly impaired accommodation (4) and appropriate spectacles enhanced academic performance (McClelland, Personal communication). Moreover, hyoscine skin patches are commonly prescribed for children with severe quadriplegic cerebral palsy, who have excessive drooling of saliva. Absence of accommodation is a common (predictable) side effect (20,21).

If accommodation is impaired, near targets are not brought into focus. This is very similar to presbyopia in the elderly. Both adults with presbyopia, and children with impaired accommodation require a near spectacle correction, and any hypermetropia needs to be fully corrected. Bifocal glasses can be given for those who have good eye movement control but separate pairs of glasses are required for those who do not. Lack of accommodation in children who are long-sighted means that the normal process of focusing to overcome the long sightedness, does not take place. In the young child this can give rise to profound amblyopia if not appropriately corrected.

Detection of impaired pupil responses to near fixation provides a method for the non-specialist to determine if accommodation is impaired and to seek appropriate refraction and spectacle correction (22).

Table 1. A question inventory designed to elicit evidence of visual field impairment and perceptual and cognitive difficulties in children with cerebral visual impairment in association with cerebral palsy. (NA = not applicable)

Questions seeking evidence of visual field impairment or impaired visual attention on one or other side	**never**	**rarely**	**sometimes**	**often**	**always**	**NA**
Does your child…						
1. miss, trip or move wheelchair over toys or obstacles on the floor?						
2. have difficulty to see the next step down?						
3. trip at the edges of pavements going up?						
4. trip at the edges of pavements going down?						
5. appear to 'get stuck' at the top of a slide/ hill?						
6. look down when crossing floor boundaries e.g. where lino meets carpet?						
7. leave food on the near or far side of their plate? If so, on which side?	near/		far			
8. leave food on the right or left side of their plate? If so, on which side?	right/		left			
Does your child…						
9. have difficulty to find the beginning of a line when reading?						
10. have difficulty to find the next word when reading?						
11. move out in front of traffic? If so, traffic from which side?	right/	left/	both			
12. bump into doorframes or partly open doors? If so, which side?	right/	left/	both			
13. miss pictures or words on one side of a page? If so, which side?	right/	left/	both			
Questions seeking evidence of impaired perception of movement	**never**	**rarely**	**sometimes**	**often**	**always**	**NA**
Does your child…						
14. have difficulty to see passing vehicles when they are in a moving car?						
15. have difficulty to see things which are moving quickly, such as small animals?						
16. avoid watching fast moving TV?						
17. choose to watch slow moving TV?						
18. have difficulty to catch a ball?						
Questions seeking evidence of difficulty handling the complexity of a visual scene						
Does your child…						
19. have difficulty to see something which is pointed out in the distance?						
20. have difficulty to find a close friend or relative who is standing in a group?						
21. have difficulty to find an item in a						

	never	rarely	sometimes	often	always	NA
supermarket e.g. finding the breakfast cereal they want?						
22. get lost in places where there is a lot to see, e.g. a crowded shop?						
23. get lost in places which are well known to them?						
24. have difficulty to locate an item of clothing in a pile of clothes?						
25. have difficulty to select any toy in a toy box?						
26. try to sit closer to the television than about 30cm?						
27. find to copy words or drawings time-consuming and difficult?						
Questions seeking evidence impairment of visually guided movement of the body and further evidence of visual field impairment	**never**	**rarely**	**sometimes**	**often**	**always**	**NA**
28. When walking, does your child hold onto your clothes, tugging down?						
29. Does your child find uneven ground difficult to walk over?						
30. Does your child bump into low furniture such as a coffee table?						
31. Is low furniture bumped into if it is moved?						
32. Does your child get angry if furniture is moved?						
33. Does your child explore floor boundaries (e.g. lino/carpet) with their foot before crossing the boundary?						
34. Does your child find inside floor boundaries difficult to cross?						
34a. If so… boundaries that are new to them?						
34b. boundaries that are well known to them?						
Questions seeking evidence of impairment of visually guided movement of the upper limbs						
35. Does your child reach incorrectly for objects, that is, do they reach beyond or around the object?						
36. When picking up an object, does your child grasp incorrectly, that is do they miss or knock the object over?						
Questions seeking evidence of impaired visual attention	**never**	**rarely**	**sometimes**	**often**	**always**	**NA**
37. Does your child find it difficult to keep to task for more than 5 minutes						
38. After being distracted does your child find it difficult to get back to what they were doing?						
39. Does your child bump into things when walking and having a conversation?						
40. Does your child miss objects which are obvious for you because they are different from their						

background and seem to ‘pop out’ e.g. a bright ball in the grass?						
Questions seeking evidence of behavioural difficulties associated with crowded environments						
41. Do rooms with a lot of clutter cause difficult behaviour?						
42. Do quiet places / open countryside cause difficult behaviour?						
43. Is behaviour in a busy supermarket or shopping centre difficult?						
44. Does your child react angrily when other restless children cause distraction?						
Questions evaluating the ability to recognise what is being looked at						
Does your child…						
45. have difficulty to recognise close relatives in real life?						
46. have difficulty to recognise close relatives from photographs?						
47. mistakenly identify strangers as people known to them?						
48. have difficulty to understanding the meaning of facial expressions?						
49. have difficulty to name common colours?						
50. have difficulty to name basic shapes such as squares, triangles and circles?						
51. have difficulty to recognise familiar objects such as the family car?						

4. Visual dysfunction due to damage to the brain

In children with CVI, damage to the input system and occipital cortex of the visual brain can impair visual acuity and contrast sensitivity, and may restrict visual fields (23,24), while damage to the higher visual processing centres causes perceptual and cognitive visual impairment. These visual systems can be damaged separately, or together, resulting in poor acuity, visual field impairment, and perceptual visual dysfunction in any combination or degree (25). Perceptual visual impairment can also occur in the context of normal visual fields and normal or near normal visual acuities (26). In children this can cause multiple problems, including impaired recognition of people and their facial expressions, shapes and objects, orientation difficulties (due to ventral stream dysfunction), and difficulty handling complex visual scenes and inaccurate visual guidance of limb movement (due to dorsal stream dysfunction) (27-30). Lower visual field impairment which may be peripheral or affect the whole lower visual field is common in children with dorsal stream dysfunction (30). The visual system may be the only part affected, or there may be associated damage to other brain structures, resulting in other developmental problems, which may also be responsible for some of the difficulties.

Children with periventricular white matter brain damage are associated with extremely low birthweight or prematurity commonly show evidence of impaired visuomotor guidance (31), impaired visual motion processing (32) and disability of carrying out tasks requiring visual closure (1) which persist long term (33) and this may be associated with cerebral palsy, in particular spastic diplegia (1,34). Disorders of the processing system, are complex, and may be elusive, particularly when there is associated cerebral palsy and this can be confounded by intellectual and developmental disorders. A good starting point in the assessment of a child with cerebral palsy is to assume that CVI is present until proved otherwise.

5. Assessment of vision in children with cerebral palsy

The approach for assessment depends upon how severely affected the child is. The spectrum of visual impairment includes profound visual impairment in the profoundly disabled child with spastic quadriplegia, (35), wide ranging and complex difficulties in the child with additional treated hydrocephalus, or peripheral lower visual field impairment associated with dorsal stream dysfunction in the mobile child with spastic diplegia who attends mainstream school (36).

Assessment of 'visual function' as carried out by the medical profession requires measurement of the thresholds, or limits, of visual function for each eye. On the other hand, assessment of 'functional vision', (evaluating how vision is optimally employed) is ideally performed in the child's own environment, with both eyes open, to determine the impact of impaired vision on daily living. Corrected visual acuities and estimation of visual fields are carried out on all children using methods appropriate to the abilities of the child (37,38). We have found that structured clinical history taking of children with damage to the brain, employing the question inventory in table 1, provides an effective and reproducible way of estimating the nature and degree of involvement of both the child's visual fields, and cognitive and perceptual visual impairment (39,40). The inventory gives a clear separation between children with visual difficulties and normal controls. Positive responses are probed and characterized by further history taking and observation of the child's visual behaviour. The questions are wide ranging and seek common difficulties for which appropriate intervention can make a significant difference for the child. An approach in which the inventory is given to the parents in advance of the appointment, and the positive responses are clarified and acted upon, has proven to be an efficient approach for our clinical service. Neuropsychological investigation can further typify and quantify the disorder, but it is important to take into account the developmental status of the child, which can preclude such investigation, or make interpretation of the results difficult with respect to visual functioning.

6. Patterns of visual impairment to look for in children with cerebral palsy

Profound visual impairment is most commonly associated with severe quadriplegic cerebral palsy, but can also, rarely, be seen in a mobile child with relatively minor problems with mobility. Children with this pattern of visual dysfunction show a range pattern of visual disability. In most cases, hypoxic ischaemic encephalopathy has affected both the occipital cortex and the basal ganglia. Total blindness is rare. Estimation of visual function by visual evoked potentials, preferential looking and optokinetic responses show very low or undetectable visual acuity, but history taking and detailed observation of the child's behaviour usually reveals evidence of 'blindsight' (35).

Adults who are blind because of cerebral damage can show evidence of relatively subconscious awareness of moving targets, lights and colors in the blind area. An unconscious ability to return facial expressions despite not being able to see them has also been described. This has been called affective blindsight (41). The brain structures which probably serve this function include extrastriate cortex, the superior colliculi and the pulvinar (42). Children, who are apparently blind according to classical clinical examination methods, may show behaviours consistent with these observations. Intermittent responsiveness to a moving target in the periphery of vision is typical. The mouth may consistently open two or three times, (but not thereafter, probably on account of fatigability of this form of visual function) for a spoon coming in from one side but not from straight ahead. Some children, in our experience may also return facial expressions, which may represent a form of affective blindsight. There may or may not be conscious awareness for the visual function in the older child (43).

6. 1. The child with spastic diplegia

In the child with spastic diplegia, periventricular white matter pathology is the most common finding on MRI imaging (34). The most common clinical picture is one of lower visual field impairment (which ranges between being extensive and being very peripheral), leading to the mobility problems outlined in table 1; as well as positive responses to questions 1-7 indicative of lower visual field loss, positive responses to the questions, which reflect deficits in dorsal stream visual processing and visual motion processing, are not infrequently seen in children with spastic diplegia in our experience.

6. 2. The child with left or right hemiplegia

Positive responses to questions 8 to 13 may lead to the discovery of homonymous hemianopic visual field defects or problems with visual attention on the side of the hemiplegia.

Table 2. Outline of the typical clinical features indicative of dorsal and ventral stream dysfunction in children (which can manifest in almost any combination and degree, both in children with no physical impairment and in those with cerebral palsy), with strategies that can assist the child

Features	Recommended strategies
Dorsal stream dysfunction	
Impaired ability to handle complex visual scenes* can cause difficulties with:	***As a rule:***
Finding a toy in a toy box.	Minimize the amount of visual information presented to the child, for example:
Finding an object on a patterned background.	Store toys separately.
Finding an item of clothing in a pile of clothes	Use plain carpets, bedspreads and decoration.
	Store clothes separately in clear compartments.
	Home:
	At home clearly organized and labeled storage systems with minimal number of items in each area can make items easier to find and locate. Both vertical and horizontal storage should be experimented with.
	Have a box, tray, plate for special items, e.g. glasses, mobile, and I pod for an older child.
	School:
	Minimize the number of items on the desk, tray, school bag
	Try transparent school bag, pencil case to make it easier to find items
	Reduce visual clutter and information beside the board, at the point in class where most of teaching is done and at the area of the class where the daily task chart is.
	Try a desk tidy.
Identifying someone in a group	Encourage child to get closer to the group.
	Identify through waving and speaking, either the child shouting for person, or the person shouting for the child depending on which system works best.
	Have a clearly identified meeting point at the end of the day when there is a crowd of adults at the school gate.
	Give additional verbal guidance to find friends in a busy play ground i.e. Jenny is playing by the steps, wearing a pink jacket
Seeing a distant object (despite adequate acuity)	Encourage the child to call or text the person they want to find.
	Share a zoom video/digital camera/ mobile phone camera view to find and record salient information.
	Training in seeking and identifying landmarks
Tendency to get lost in crowded locations.	Visit shops when they are quiet.
Distress in busy shops and crowded places	Determine whether covering surrounding text improves reading ability.
Reading when presented with a lot of information on the page/ board.	Present small amount of *san serif* clear well separated text, with small amounts of text on each page (matched to the child's abilities). Wear long sighted correction.
Lower visual field impairment / Impaired visually guided movement (optic ataxia)	
Upper limbs: Inaccurate visually guided reach that may	Practice eye hand coordination skills, using games, Wii , Play station

be compensated for by reaching beyond an object, or placing whole hand over object then gathering it up. Lower limbs: Feeling with the foot for the height of the ground ahead at floor boundaries. Difficulty walking over uneven surfaces (Despite full visual field, and looking down.)	Provision of tactile guides to the height of the ground ahead. For example pushing a toy pram or holding on to the belt pocket or elbow of an accompanying person. Highlight floor boundaries, edges of steps and surface changes. Provide additional assistance when going over uneven ground, verbal reminders, physical support e.g. straight arm to hold, banister or railing at the correct height. Contrast colours of different floor surface. Try plain floor surface. Keep floor surfaces free from clutter, have organised storage systems. i.e. no bags or shoes left on floor. Try white trainers to increase visibility of feet. ***School*** Keep a clutter free pathway from door to child's desk, from child's desk to teacher's desk and to white board or floor sitting area.
Hemianopia or lack of attention on one side	Avoid changes to classroom layout when possible and if changes are made involving the child. Give permission to use stairs before or after the rush and/ or encourage child to wait at the end of line when using stairs. Position so that target of interest (e.g. the teacher) is off centre to the sighted side. For lack of attention on one side, place task work on the work station off centre to the sighted side, and teach from that side.
Impaired attention Difficulty multi-tasking Difficulty "seeing" when talking at the same time, which may cause a child to trip or bump into obstacles. Frustration if distracted	Only make one demand at a time – listen, or write, or walk or talk, understand that watching a face whilst talking/ or listening can be difficult and encourage child to only attempt one task at a time. Minimize distractions around the work area Give short tasks Give short periods of focus time Vary positioning of tasks i.e. sitting, standing, and provide movement breaks if required Break home work task into short blocks Complete one short piece of work at a time Alternate work tasks with fun time Use a visual or sound timer Have a quiet work space without distractions i.e. no music, TV, or siblings while working. Minimize distractions in work area or provide a quiet work space in the classroom. Allow a fidget object in hands to help concentration when listening i.e. giant paper clip, rubber, Blu Tac. Limit distraction by minimizing background clutter and background activity.

VENTRAL STREAM DYSFUNCTION	
Impaired recognition	
Difficulty to recognise people and photographs	Family and friends introduce themselves and wear consistent identifiers. Child requires information and training to recognise identifiers.
Difficulty to recognise shapes and objects	Encourage training in tactile recognition as well as visual.
Impaired orientation	
Tendency to easily get lost in known locations.	Training and practice in orientation using memory maps, instruction following and meeting points.

6. 3. The child with dyskinetic cerebral palsy

It has been our experience, after having an index case, that impairment of accommodation is common in children with dyskinetic cerebral palsy (44). This may be accompanied by disorders of gaze, including impaired up-gaze. Near acuity is poorer than distance acuity, but this is corrected by an additional 1.5 - 3 dioptre spectacle correction. Dynamic retinoscopy confirms the diagnosis.

7. Strategies to assist children with visual difficulties associated with cerebral palsy

Identification of any visual problems described in table 1 naturally leads to devise appropriate strategies. Table 2 provides an outline of this approach, which parents and teachers have, in our experience, found to be helpful. The act of identifying and explaining the nature and degree of visual problems makes a significant difference for each child. Not only appropriate action can be taken, but also there can be significant change in attitude of parents, teachers and carers at home and school. The child is no longer thought to be clumsy or badly behaved; and negative approaches are replaced by positive supportive practice. The change to a positive supportive ambience, from one which was negative and critical, can change a child's life. "Can't you see that toy in front of you?" becomes "Your toy is just there".

8. Conclusion

Cerebral visual impairment is a common association with cerebral palsy. It contributes to impaired visual guidance of movement, and a wide range of visual disabilities which can be misconstrued. Characterization of each child's visual dysfunction allows for the management plan to take into account the resultant difficulties in a constructive and structured manner.

The recently identified plasticity of the visual system and the finding that training can enhance visual function leads to the concept that training programmes matched to the nature and degree of visual dysfunction are likely to be developed in the future. A textbook which provides more detailed information concerning cerebral visual impairment and its management has recently become available (45).

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