

**Types of impaired vision in children related to damage
to the brain, and approaches towards their management**

Gordon N Dutton

Paediatric Ophthalmologist

Emeritus Professor of Visual Science

Glasgow Caledonian University

Cowcaddens Road

Glasgow G4 0BA

UK

**(Paper presented at the Proceedings of the South Pacific Educators for the
Visually Impaired. January 2013.)**

Abstract

Cerebral visual impairment is complex. Affected children commonly have refractive errors and need spectacles. Impaired control of eye focusing can cause blurring of vision unless corrected. Damaged visual pathways can cause lack of acuity and / or disordered visual fields. Abnormal eye movement control impairs capture of rapidly moving information. Disturbed ventral stream processing can impair recognition and / or route finding. Dorsal stream damage limits analysis of complex visual scenes and visual guidance of movement. Children with impaired vision from damage to the brain need structured history taking and assessment encompassing all elements of vision, to find and characterise all visual limitations. Educational approaches ensure that dimensions, contrast and location of material shown, fall within visual limitations. Each child's unique perceptual constraints are also identified, catered for and worked within.

Key words: Prematurity. Cerebral visual impairment. Periventricular white matter. Neuroplasticity. Habilitation.

How does the brain see?

Cerebral visual impairment (CVI) is the commonest cause of visual impairment in children in the developed world. A large part of the brain is devoted to seeing, so it is not surprising that conditions that affect the structure or function of the brain can interfere with vision (Dutton and Bax 2010).

At a basic level, the front of the brain 'thinks', 'understands' and 'behaves'; the middle 'feels and moves the body, and hears and understands language'; while the back 'sees'.

The picture made by the eyes is converted into electrical signals and the visual information is sent to the back of the brain where it is sorted out in different ways:

1. Processing the picture

First, at the back of the brain (the visual cortex in the occipital lobes) the detail of the picture is sorted (measured as visual acuity); colour is perceived; shades of grey are differentiated (measured as contrast sensitivity); a wide view is obtained that is clear in the centre and more blurred to the sides (measured as the visual fields), and the speed and direction of moving images are processed.

2. Guiding our movement and seeing lots of things at once

Second, at the top of the brain at the back (the posterior parietal lobes, connected to the occipital lobes by the dorsal stream pathways)), there is an amazing system. The visual information is carried there from the occipital lobes by a connecting pathway, called the dorsal stream. For every moment of our waking lives, this area of the brain uses the incoming moving image data to guide our movements through our surrounding environment (Milner and Goodale 2006).

Reach out and pick something up. You 'know' that the object that you see is out in front of you, but the picture that you perceive isn't really, is it? It's inside your brain! What is remarkable is that the picture out there in front of you is so compelling that you believe that it is there, but you are actually moving through the picture in your brain, fully expecting that it will coincide with the reality of the object you are about to pick up, which indeed it does. This counterintuitive idea can take a while to get one's head round, but it gets even more complicated. The whole process takes place completely unconsciously!

Some people who have gone blind in both eyes because of damage to the back of the brain, which spares the movement seeing centre, are able to accurately move through the world that they do not consciously see (Goodale and Milner 2004). Functional scans of the brain show that they do this with their intact posterior parietal lobes. "But wait a minute" you may be thinking, "I know

what I can see; and I was able to choose and know what I picked up." Yes, but the conscious seeing and knowing is performed separately by the closely interlinked but distinct lower part of the brain at the back, the temporal lobes, which are connected to the visual cortex in the occipital lobes by another pathway, called the ventral stream.

To move rapidly and efficiently through our visual surroundings without bumping into people or obstacles, we must populate the three dimensional moving virtual imagery in our minds, with all the surrounding items, and locate them accurately in the three dimensional virtual reality headset of our minds, which we assume to be coincident with reality! It is this virtual framework that underpins visual interest and the ability to give attention to elements in the visual scene. It also allows us to carry out visual search. This remarkable unconscious framework is connected to the frontal lobes of the brain that make the executive choices about where to move and what to pick up, and bring about the accompanying rapid eye movements that we use to look in more detail at items of interest. It is also connected to the temporal lobes so that we can know and recognise what we are looking at.

3. Recognition

The **third** component of the visual brain is to be found in the temporal lobes where conscious recognition takes place.

You recognise a friend in a group of other people whom you don't know. To do this, you have a memory store of all the people you do know. In came the picture of the group of people. The pictures of all their faces were compared with your memory store and there was one match, - your friend. The two of you find your way to a restaurant, and recognise all the objects, the cutlery and the crockery. The temporal lobes do this for you by comparing this incoming visual information of surrounding geography, people and items, with your comprehensive visual memory store that has been built up since the day you were born. If there is no match, then the route, item, person or shape cannot be identified and they are 'cognised' for the first time, but when there is a match they are 're'-cognised. What extraordinary computing, yet we take it for granted!

The two 'higher' visual analysis systems, the dorsal and ventral streams, are blended so well that they work together in harmony, but of course we are unaware of all the unconscious visual workings of the brain, and therefore believe, wrongly, that we have conscious awareness of everything that our brains are seeing and handling.

In addition to 'higher' visual functioning, there are myriad automatic visual systems. Examples include:

- A. The time clock of when we go to sleep and wake up is driven by our perception of light and dark. (Children who are totally blind can live a 26 hour day because they lack this visual correction factor.)
- B. At the top of the midbrain at the back and in the thalamus, there is an automatic visual system, that is alert to visual hazards and helps us to reflexly and automatically avoid them.
- C. Our eyes are perfectly internally stabilised by our balance systems that even make compensatory eye movements for our heartbeat let alone for moving through 3D space.
- D. Our eyes need to slow down and stop each time we look at something. The cerebellum contributes to this.

So how does this information help in finding out whether a child has cerebral visual impairment?

Even with the right glasses, each child with cerebral visual impairment can have a unique range of visual difficulties, due to any of the processes outlined not functioning perfectly, and this can occur in any combination or severity.

Spectacles. What are they for and when are they needed in children with cerebral visual impairment?

Many children with cerebral visual impairment wear glasses. Spectacles are worn for different reasons.

Short sightedness (myopia) is common in children born prematurely. A short-sighted eye is a large eye and the picture is brought into focus in front of the retina. Glasses with concave lenses are used to move the image backwards onto the retina. The glasses bring the distant world into focus. They make the eyes look smaller and one can see that the side of the child's face viewed through the glasses appears to be displaced inwards. Without glasses the child can see for near, but distant objects are out of focus, hence the term short sight. For some children with reduced clarity of vision due additional damage to the visual system, this means that the printed page can be seen better without glasses. The short sight means that the child can see the page when held close to the eyes. The magnification gained by this proximity gives an effect equivalent to a magnifying glass.

Long sightedness (hypermetropia) occurs when the eye is small. The picture is blurred because it hits the retina before it has been brought into focus. In most children with long sightedness the focusing system of the eye compensates and brings the picture into focus, but in many with damage to the brain, long sightedness needs to be corrected with glasses because the automatic focusing system (accommodation) may not work so well and even small amounts of long sightedness need to be corrected for (Saunders et al 2010). This problem can affect over half of such children in special schools, and a small correction for what would otherwise be a 'normal' amount of long sightedness can make a

significant difference to the child's ability to see, as well as their academic performance. In these children, the pupils of the eyes do not become smaller in the way that they should when looking at a close item. This observation provides a way of identifying those affected (Saunders et al 2008). Some affected children need an additional bifocal correction or a separate pair of glasses for reading. Long sightedness can also be responsible for making the eyes squint by turning in and when glasses are worn the squint is reduced or eliminated. In children who are long sighted the spectacles magnify the eyes and ideally need to be worn most of the time.

Astigmatism occurs when the front of the eye has a very slight rugby ball shape. The lens of the glass is shaped in a reciprocal way to compensate for this shape anomaly.

Children with reduced vision due to damage to the brain commonly remove their glasses even when these definitely improve vision. One possible reason is visual fatigue. The clear picture requires more processing power than the blurred picture and perhaps it is more comfortable to have a blurred picture when tired, because it is less complicated and easier to handle. This is of course only a working hypothesis, but when children remove their glasses after working hard for a while, they may be giving their visual systems a well deserved rest.

In summary, short-sightedness is corrected by glasses that make the eyes look smaller, and children may remove their glasses to gain magnification by holding things up close. Long-sightedness is corrected by glasses that make the eyes look bigger, and children may remove their glasses to get a more comfortable blurry picture for a while.

Visual Thresholds

We are all limited by our vision but because we 'know' our vision to be normal, but we are not aware of these limitations. Telescopes and magnifying glasses allow us to see things that are otherwise too small to visualise. This highlights the limitations of the clarity of our own vision. Society chooses to present information in a way that the majority of people can see clearly, by ensuring that it falls within these 'normal' limitations, but those with reduced vision are not able to always see within these limits and the information is not accessible, nor is it visible or even known about.

Children with poor vision from an early age also 'know' their vision to be normal and just like fully sighted people, they do not know what it is they do not see and they are not aware of their visual limitations. Unless we know in detail what these limitations are for each child, information that is not seen will be presented. Failure of the child to respond can easily be misinterpreted as lack

of ability or even stubbornness. It is very expensive to teach children with damage to the brain and the cost implications alone of using educational material and approaches that are not actually seen by the children, are considerable, never mind the adverse impact upon the child's learning, which can well be misinterpreted as being due to impaired intellect. It is therefore important to measure functional vision as accurately as possible, so as to get inside the mind of the child, so as to understand as fully as possible what is seen and what is not seen, and then to work inside all of these limitations or thresholds.

Clarity of vision

Look at this printed page and move it back until you can only just read it. You are now looking at the smallest text you can see when it is printed in black against a white background. This is your visual acuity or the limit of your clarity of vision.

Now try and read the text as fast as you can when held at this distance (and don't cheat by getting closer to it). You will find that it is something of a strain and that you quickly fatigue and get fed up with the task. Compare the time it took you to read the passage with the time it takes you to read the same text when held at your normal reading distance. It is much quicker at the normal reading distance and much less tiring. This is because the text is now big

enough to be seen clearly and comfortably.

The letter charts used to measure clarity of vision measure visual acuity. This is the main test used by eye care specialists to make a diagnosis of reduced central vision and to measure the smallest clear black target that can be seen against a white background. However, this is not a measure of functional vision (which is recorded with both eyes open), it is a measurement taken for medical reasons in order to help make a diagnosis or to provide information required for follow up. For a child who has reduced vision, what one wants to know is the size of target that can be seen with ease at maximum speed, this is the functional visual acuity.

Educational information needs to be well within the limit set by the binocular functional visual acuity so that it can be easily seen throughout the day even when the child is tired.

Colour and contrast

Imagine the rainbow. Red, orange, yellow, green, blue and purple all blending into one another. Purple can also blend into red. These colours can be arrayed in a circle in such a way that they blend with each other. This is known as the colour circle. If you take a can of grey paint and add red colouring little by little, the paint will gradually become redder until it can get no more red when it

becomes saturated. One can now imagine a central grey circle with a surrounding rainbow of colour that become progressively more colourful to yield the primary colours on the outside. Imagine now that the grey centre becomes progressively whiter vertically upwards and progressively blacker vertically downwards. This is what is known as the grey scale. Finally imagine the red becoming a progressively lighter shade of pink until it blends into the apex of white at the top, and becoming progressively browner as it blends into a black apex at the bottom. The same idea can now be applied for all the colours. This three dimensional imaginary concept is known as the colour solid, being made up of two circular cones base to base, in which one can conceive of every imaginable colour, hue or shade in its own location within the 3D construct. Black and white are the furthest apart, and show the greatest contrast while, for example, blue and slightly lighter blue are close together and show very little contrast.

In relative darkness it is easy to mix up such colours as green and brown, but black and white can still be differentiated from each other, as can very light blue and navy blue. Many causes of visual impairment cause difficulties in differentiating contrast and colour. This of course imposes limitations on what can and cannot be seen. As a rule of thumb, the further apart two colours are across or within the colour solid, the more likely they are to be seen as being different.

Contrast perception can be markedly impaired in children with cerebral visual impairment (Good et al 2012), and needs to be measured, but the degree to which brain damage impairs colour vision and contrast perception in affected children is not known. As a general rule colour perception is maintained remarkably well in such children but from a practical point of view it is worth ensuring that pictures and toys are bright and clear, and that most colour boundaries contrast well. For example a picture of a dark green frog against lighter green grass may not be seen because of the low colour contrast, but the same frog portrayed on yellow sand would be much more obvious. Information is often photocopied and re-photocopied. This results in grey text against a grey background. The contrast is therefore reduced.

The visual world is made up of myriad colours in all sorts of juxtapositions. Visual impairment can degrade colour boundaries of low contrast, while not affecting boundaries with more contrasting colours and shades. This means that some things are seen and others are not and it is important to be vigilant to look out for what is seen when not expected, and what is not seen when it is expected. It is by making mental notes about these observations while working or just being with the child, that one can build up a mental picture of the colour boundaries that can be seen and those that cannot.

Visual fields

The visual field is the area that one can see at any one time. The nerve fibres that run from the eyes to the brain are arranged in a very organised manner. This means that when damage takes place, well-recognised patterns of visual field impairment can occur, and different patterns of impairment have different functional consequences.

Hemianopia

The wiring diagram of the brain is such that occipital lobe on the right is responsible for seeing on the left side of the visual scene (for both eyes), and the occipital lobe on the left is responsible for seeing the right side of the visual scene. Occipital lobe damage can affect one side or the other and gives rise to left or right-sided lack of vision.

The lack of vision in hemianopia can be thought of in the same way as the world behind you. It is not seen. In a good proportion of children affected from birth, however, some visual function may be retained such as subconscious perception of movement, so that an affected child may not appear to see on one side but is able to walk through a crowd without bumping into anyone.

Damage to the top of the brain at the back on one side or the other (the posterior parietal lobes), can cause lack of attention on the side opposite to the

damage. As a rule of thumb lack of attention on the left side of the body (due to right parietal damage) tends to be more marked than lack of attention on the right side (due to left parietal damage).

Hemianopia can be compensated for by head or eye movements to the affected side. This allows things on the blind side to be seen. In contrast, lack of attention on one side is remarkably, not compensated for in this way and the child needs to rotate the body towards the more poorly attended side to become aware of what is present on that side. Affected children often compensate by turning their chair slightly when sitting at a table. In this way both the knife and fork can be seen. (Children with hemiplegia need to be observed for evidence of lack of attention on the affected side.)

Some more severely affected children can have both hemianopia and hemi-inattention. They do not see large moving targets on one side; head and eye movements do not compensate, and body rotation, or displacement of materials to the 'good side' is required.

From a practical point of view there are a number of issues that need to be considered when looking after a child with hemianopia or hemi-inattention.

Eating food can be a problem because food can be left on the side of the

plate, on the same side as the hemianopia or the lack of attention. When this is recognised, turning the plate round so that the remaining food comes into view can be effective. A policy of putting favourite food on the hemianopic or poorly attended side of the plate can help a child to develop strategies for exploration, because one never knows what pleasant surprises may be waiting round the corner.

Communication with a child with hemianopia or lack of attention needs to take into account that someone sitting or approaching from the visually impaired side may not be seen.

Mobility can be impaired due to hemianopia or lack of attention, with the child bumping into things and people on the affected side.

Crossing roads is an important issue. Oncoming traffic can be missed, particularly if it is small and silent like a bicycle. When looking to the affected side the head and eyes need to be turned fully, and for those with lack of attention on one side, rotation of the whole body to the affected side may be required to become aware oncoming traffic. This is perhaps best taught by example, particularly with the young child.

The position in the classroom needs to be selected so that the subject of interest

is either straight ahead or on the unaffected side. If the child is sitting so that the teacher for example is on the affected side it may be difficult to attract the child's attention.

Access to information can be restricted by hemianopia or inattention on one side because data presented on the affected side may be missed.

Reading may require special attention, with left and right hemianopia (or lack of attention on one side) having different implications in some children, especially when the onset has been later in childhood. As the eyes scan across the text the hemianopia moves with the eyes. For lack of vision on the right side, each new word can jump into view and may not be anticipated because it cannot be seen when looking straight ahead, but once the end of the line has been reached the left hand end of the next line down is seen and the eyes can jump to the beginning. For left hemianopia, on the other hand, as the text is read from left to right the text on the left progressively disappears, so that it can be difficult to find the beginning of the next line. It can be helpful to have a system of progressively moving a finger down the left hand margin. Alternative approaches of reading text either vertically or obliquely can prove very helpful for some children, particularly those who have developed their visual impairment due to damage to the brain after having learned to read.

Quadrantic visual field loss

Damage to the brain can cause lack of vision in any of the four quadrants of the visual field, affecting both eyes equally. While quadrantic visual field loss of this nature is less of a problem than hemianopia it can still cause significant problems in any of the areas outlined above,

Lower visual field impairment

The visual pathways that lead from the eyes to the brain, run very close to the water spaces in the brain called the lateral ventricles. In particular it is the fibres that serve the lower field of vision in both eyes and run over the top of the water spaces in the brain (the lateral ventricles) and lying closest to them that are most likely to be damaged. The commonest scenario is the child who has difficulty moving her legs due to spastic diplegia, who also has lower visual field impairment. When looking straight ahead she is unable to see the ground in front of her and when walking over irregular ground she has to walk with her head turned down to check whether there are any obstacles or pot-holes. On top of all that, there may also be problems using vision to locate the feet accurately.

Lower visual field defects can vary and range from being complete, so that none of the ground ahead is visible, to being relatively minor, so that only the ground one to two metres ahead is not seen. It is worth simulating a lower visual

field defect for oneself by holding a piece of card below one's eyes so that the ground immediately ahead is not seen when looking straight ahead. It is remarkable how much we take for granted. When one can't see where one's feet are treading, it is quite disabling. A systematic approach, which encourages the child to regularly look at the ground ahead to check for safety can prove helpful.

Children with impaired walking due to spastic diplegia and lower visual field impairment can particularly enjoy horse or pony riding. The horse provides mobility over rough ground; it can see where it is going, and the training in balance is also helpful. Horse riding can afford a new-found freedom for such children.

Visual field constriction

There is a range of disorders of vision due to damage to the brain, which are accompanied by constriction of the visual field. A central island of vision is present but peripheral vision is restricted. The commonest cause of apparent constriction of the visual field is difficulty attending to a lot of information at the same time, this is usually due to damage to the dorsal stream on both sides, resulting in inability to see many items at once and inevitably impairing visual search. This gives the impression that the visual field is narrow but when the visual scene is made less complicated, the apparent lack of attention to a

target in the peripheral visual field is much less evident or even no longer present.

Central visual field impairment

If the visual acuity is reduced then there is a reduction in central visual function overall, which in turn represents a central visual field impairment. In such children the more peripheral visual field may provide more useful vision and the affected child can appear to look past what he or she is looking at, when in fact the child has chosen the head and eye position that gives the clearest picture. This is called eccentric viewing.

Combinations of visual field disorders

Poor central vision commonly accompanies hemianopia, and lower visual field impairment can also accompany hemianopia so that vision is only present in one upper outer quadrant of the visual field for both eyes. Under these circumstances all of the difficulties outlined above can be compounded because of the greater restriction of vision.

Associated cognitive visual problems

Problems of recognition and orientation are quite frequently associated with hemianopia.

Limitations imposed by disorders of eye movement

Disorders of eye movement that impair vision can be divided into squint, impaired tracking, and to and fro oscillation of the eyes, or nystagmus.

Squint or strabismus is a condition in which the eyes are not aligned correctly and one of the eyes is turned in, out, up or down. The brain adapts to squint in children by ignoring the image formed by the squinting eye. Our ability to see in three dimensions relies on the differences between the two pictures presented by the eyes. These differences are interpreted as a sense of depth. If you close one eye as you reach for something you will find that your reach becomes slightly less accurate. If you play a racket sport with one eye closed you will find it more difficult to hit the ball because the two eyes act in harmony to allow you to judge speed and distance in 3D in real time. If one eye is squinting the facility for this form of 3D vision, or stereopsis is absent.

Many children with cerebral palsy have difficulty controlling the movement of their eyes. Our eye movements can be divided into two types, fast and slow. Fast eye movements are used to look from one object of interest to another, while slow eye movements are used to track a moving target. Either or both can be impaired. In addition to degrading the ability to see detail on moving targets, there can be difficulty in accessing information on static targets as well. In order to read we have to move our eyes in a very regular way. The eyes make

four or five jerking movements to the right as we read a line and at the end of the line they jump back to the beginning of the next line. In children with impaired tracking, these movements are inaccurate and reading is difficult. It is not surprising that some children appear to miss words out, or jump to the wrong line when reading. An approach that recognises this is to enlarge the text. This means that even if the successive eye movements are irregular, the next word is seen because the enlarged words help compensate for the inaccuracy of the eye movements.

To and fro movements of the eyes sometimes accompany cerebral palsy and may also be seen in children with cerebral visual impairment without cerebral palsy. One might expect that affected children would see everything oscillating to and fro, but they do not, because the brain smooths out the picture. The outcome is that the visual acuity is diminished because of the 'camera shake'. Many children discover that they get clearer vision if they hold their eyes in the position in which the eye movements are least. This results in the child adopting a head posture, particularly when concentrating on small print for example. Head posturing can be lessened in many children by enlarging the print so that there is no longer a need for the child to enhance vision in this way to the same degree.

Seeing movement

We take for granted our ability see and recognise moving targets. Not only can we see the moving object, and work out its speed and direction, but we can see and interpret its detail and recognise people and animals by the character of their movement. We all, however, recognise that when an object progressively speeds up, detail is lost; the object then becomes blurred and then invisible. As propeller blades rotate faster they become invisible, and we take it for granted that a bullet emerging from a gun barrel is invisible. In both cases the moving object is not seen because it is moving faster than the detection system in our brains can cope with.

The 'computing system' in the brain, which enables us to cope with perception of movement, is complex and it is perhaps not surprising that it may not work so well in some children with brain damage. There are two types of condition that impair the ability to see moving targets, namely impaired tracking and impaired movement perception.

In children in whom there is damage to the eye movement systems there can be difficulty in tracking moving objects. Careful observation shows that such children may be able to compensate by moving the head to follow a moving target if it is moving slowly enough, but if the target moves quickly it may be missed because the eyes cannot lock on to, and follow the moving target.

Impaired movement perception due to brain damage (dyskinetopsia) is rare. There is a small sector of the brain, at the back of the brain on both sides, which is responsible for seeing movement (the middle temporal lobes). In the majority of children with brain damage this part of the brain is preserved, and they are able to see moving targets even if brain damage is severe. However, in a minority it is this sector of brain tissue that may have been selectively damaged. Although an affected individual can see static targets, moving objects may be invisible unless they are moving slowly (Guzzetta et al 2009). This problem tends to be permanent and it is clearly very important to recognise it. Inability to recognise an object by the character of its movement is another related deficit (Pavlova et al 2003).

Children with impaired movement perception often choose to watch television programmes in which there is limited movement, such as the weather man or the news reader, but not surprisingly they have little or no interest in fast moving programmes such as cartoons.

A educational approach that takes impaired tracking or dyskinetopsia into account, involves slow movement and gesture, and avoids material such as videos or DVDs that have a lot of movement.

Moving through the 3D world

Many children with damage to the brain causing visual impairment, have profound problems bringing about accurate movement through visual space because the pathway in the brain that passes the details of the picture to the part of the brain responsible for using vision to guide movement, the dorsal stream, has been damaged. This condition, which is not uncommon, but is poorly recognised, is called optic ataxia and may be a significant element of cerebral palsy.

In some children it can be visual guidance of the leg movement through 3D space that is impaired. In others it can be guidance of the arms and hands that is the problem; or it may be visual guidance of both the arms and legs that is impaired (Dutton et al 2004). These difficulties can compound disability due to weakness and / or spasticity, or they may simply be visual in origin. Children with these difficulties often use touch to compensate, running their thumbs along the piano for example, to know where the keyboard is, while using the fingers to play. An affected child may not be able to place pegs into a board unless the board is touching a part of his body so that it can be localised.

One situation that is common is the child who has difficulty in knowing whether a line in the floor is a step or not. A boundary between linoleum and carpet for example, has to be carefully explored with the foot (or even the hands) before it

can be crossed (Dutton et al 2004). Another typical problem is difficulty negotiating steps and kerbs. Going up stairs is often easier but going down stairs is a particular problem because it is very difficult to estimate the depth of each step. The same applies to kerbs. Typically the foot is lifted too high and it may be lifted too early before coming to the step.

Other children can have problems accurately reaching for things and manipulating them. It can of course be difficult to work out whether the problem relate to weakness, or to poor coordination, or are due to visual dysfunction, but the typical picture is one in which the reach is intermittently accurate. The hand is not accurately pre-configured to the shape or to the orientation of what is about to be picked up. The gap between the fingers can be too wide, or the outstretched hand may be brought down onto the object, or the hand reaches out beyond the object to gather it up.

Practical approaches to these problems require repeated practise.

Supplementing visual guidance with touch, in a manner akin to blind techniques, may be naturally adopted, or it may need training. Quiet soft play areas that provide a stimulating opportunity to learn to move through 3D space without injury can be very helpful, both in providing the opportunity to learn skills and in helping confidence to develop.

Crowding and complexity

If one opens too many programmes in a computer to run at the same time the computer gets slower, and if one more programme is opened the computer stops working. This is because there isn't enough active memory to cope with all the tasks that need to be done at the same time. Our minds function in a similar way. Watching television whilst doing homework usually means that it takes longer to do the homework. If there is a conversation going on in the room at the same time, the homework may never get done because all the 'programmes' cannot be held open at once.

The visual system has to handle a very large amount of information at the same time and it does so by means of a complex system in the posterior parietal lobes. All the incoming information is processed simultaneously and unconsciously, but the conscious mind cannot cope with it all, so there is a second selection system located in the frontal lobes that selects out which information to attend to at any one time, and allows the rest to be ignored until it is chosen later for future attention.

Damage to the posterior parietal lobes or to the pathways linking them to the visual system, the dorsal stream, means that the mind cannot cope with a lot of information at the same time. Children with such problems show a number of different features related to the complexity of the visual scene. Both the

background and the foreground can be detailed and difficult to fully appreciate. Young children can find it difficult to locate a toy when it is on a patterned carpet but have much less difficulty finding the same toy on a plain carpet. If the same toy is in amongst other toys on a plain carpet it may also not be possible to find it.

The practical approach is to regularly investigate how clear the background and the foreground have to be in order to allow the child to function optimally. Older children who are learning to read may only be able to access a small number of words at the same time. The approach to take in this situation is analogous to reading Braille, in which the information is broken down and presented sequentially. For example when learning to read, a computer can be used to show one word at a time. When the condition is less severe, enlargement of text reduces crowding and can help significantly. Magnifying aids can also help because they too diminish crowding. A spectacle correction to correct long sightedness that magnifies a little, may help. Another approach to consider, is to try using a card with a slot cut in it (a typoscope), to mask surrounding text. For arithmetic, it can help to use squared paper to locate numbers (ensuring the the lines of the squares are visible), and to present fewer problems per page.

Impaired simultaneous perception (simultanagnosia) can give the impression

that the child has tunnel vision because it is not possible to attend to a visual stimulus at the side at the same time as attending to something of interest in the centre of the field of vision. The visual acuity may be normal for single letters but reduced for words. This is called crowding and is brought about because the more information is present, the bigger it has to be to minimise the crowding of the central visual scene.

Gradual spontaneous improvement over a number of years takes place in the majority of children, and the ability to handle increasing amounts of visual information gradually improves. This means that the condition needs to be kept under regular review so that the educational approaches employed are matched to ability as the child grows up.

Recognition

The human brain is designed so that we can rapidly see, know and understand what we are looking at. When a baby is born and looks around for the first time, the brain, which is a remarkably active self-programming computer, is turned on. The brain of the newborn infant is rather like a brand new library without many books in it. There are however a number of rooms in the library destined for book collections about different subjects. The room for face recognition already has one or two books in it. The newborn infant will spend more time looking at a face than a jumbled pattern of two eyes a nose and a mouth, and

as each new face is seen, the picture is stored for subsequent recognition. Seeing the same face many times means that the young child comes to recognise close family members. As time goes by and the baby explores, the information about what is experienced is given meaning and is progressively stored. If there is an impaired visual input then the visual information which is stored can only be as good as the quality of the input that is provided. The mind can only learn to see as well as the information it is provided with. Where clarity of visual input is low, the young child compensates by getting closer to view things. The magnification obtained by proximity compensates for the impaired input. In contrast, when there is brain damage present, the 'computing units' responsible for knowing and understanding what is seen may be dysfunctional, leading to poor recognition.

Recognition of people

When you walk down a busy street and recognise and greet a friend, the amount of computing being done by your brain is phenomenal. The act of not recognising someone needs a lot of processing. Each person is compared with the whole stored image bank of the hundreds of people you already know, and an almost instant conclusion is reached that you haven't met that person before. When you come to the person you do know, a match is made and you are able to greet your friend.

During your subsequent conversation you are able to respond to a wide range of nuances of facial expression and reciprocate with appropriate facial expressions of your own. Although we take facial expression for granted, this too needs a lot of computing power in the brain.

Children who have poor vision due to brain damage can have impairment of both face recognition and the ability to interpret facial expression (Fazzi et al 2007; Ortibus et al 2011). For obvious reasons, it is very important to recognise these disabilities. An inability or disability in recognising one's friends is socially disabling. If this is compounded by not being able to react appropriately to facial expressions, a significant degree of alienation can result. When teaching a child with these problems one has to be aware that one is recognised by the sound of one's voice and that the language conveyed by facial expression may not be evident. The voice therefore needs to convey information as well as clear explanation of emotion and feeling. (This is also true for all children with insufficient visual acuity to see faces, for whom it is essential to know the face and facial expression recognition distance.)

It is important to recognise that impaired face and facial expression recognition, can potentially be misinterpreted as the impaired theory of mind typifying autistic spectrum disorder.

Recognition of shape and form

In order to recognise the differences between different types of car, the brain has to do the same job as it does for faces but a different part of the brain is used. This means that brain damage can result in problems in differentiating shapes from one another, but with an intact ability to recognise faces. This can be relevant to maths for example where such a child may have numeric skills but be unable to comprehend geometry by means of vision. (Feeling geometric shapes, or strings representing these shapes, and later envisioning their haptic characteristics can help compensate.)

Damage to the temporal lobes can rarely impair the ability to read text resulting in alexia (inability to read) or dyslexia (selective impairment in reading in the context of normal intelligence in other aspects of intellectual function). The part of the brain responsible for converting the written word into language comprises the language centre, which is on the left side of the brain in most people. If there is damage to the back of the brain on the left combined with damage to the pathway between the back of the brain on the right to the language centre, alexia is the result. The damage on the left causes inability to see on the right side or hemianopia. There is intact visual function in the right brain, but because the pathway (the posterior corpus callosum) that conveys this information to the reading centre is damaged, text information cannot be interpreted linguistically (O'Hare et al 1998). There is some evidence that

phonetic reading is particularly impaired in these individuals and that the 'look and say' method of reading may provide a means of teaching individuals with this rare disorder.

Orientation

Orientation is not truly a visual skill because people with no vision can be fully orientated by virtue of their other senses. However, in general, vision and visual memory play a large part. We need to be orientated to find our way from one place to another. The same skills are needed to know where to find things in cupboards and drawers, both at home and at school, and orientation is needed to know where one has put things down. If the part of the brain used for orientation is not functioning well there may be problems and difficulties, both on the large scale of finding one's way around, and on the small scale within the home and at school (Dutton et al 2004).

Like the skill of face recognition, orientation requires an ability to retain a store of information, which is compared with the current scene. If there is a match one is orientated if not the new scene needs to be learned and memorised for future reference.

The part of the brain used for finding one's way around is called is close to the part for recognising faces (the fusiform gyrus of the left temporal lobe) and close

to the part for seeing on the left hand side. This means that children with impaired orientation may or may not have additional problems recognising people, and seeing on the left hand side.

Orientation when outside

It can be difficult to know whether a child with brain damage has difficulty finding his way around outside because such children rarely have the opportunity to get out and about on their own. There can therefore be two factors leading to problems route finding, an intrinsic disability, and lack of opportunity to learn the strategies that come naturally to children who are given their independence. When possible the child should be asked to be the guide and take the lead.

There are a number of approaches that can be helpful for people who have difficulties finding their way about.

- Talking about where one is going in a consistent way for all important routes, helps each route to be remembered.
- Looking out for landmarks and talking about them.
- Learning where the sun is at different times of day and learning how to use the sun as a reference point so that one doesn't lose one's sense of direction.
- Writing short songs or poems about important routes can prove very

helpful to some.

- Playing hide and seek.
- Getting out and about regularly

Orientation when inside

Have you ever had difficulty remembering which drawer or cupboard something is in? Imagine what it is like to have this as a permanent problem, but you know that you are normal because you have always had the problem and you do not know what it is to have the skill. You develop a system of leaving things in specific locations, which you have spent ages remembering. Then someone moves them! Imagine how frustrating this is. It takes ages to find them despite a huge amount of effort and then you are told that you're stupid if you can't find things. Not surprisingly your frustration mounts. You then develop a system of marking each drawer and each cupboard and your mum comes into the room, gets angry that the furniture has been defaced and removes all your carefully designed labels. Your frustration boils over and you are thought to have behavioural problems! This is a true story of a child with problems due to previously undiagnosed problems with orientation. As soon as the problem was recognised and everyone was informed of the cause and nature of the difficulty, the drawers were re-labelled, the position of everything was respected and all the 'behavioural problems' disappeared.

Affected children have tremendous problems when they change schools, and need intensive orientation training in such new environments. The degree of difficulty can vary from occasionally getting lost in school, to never being able to find the classroom without help. Identifiers designed and put up by the child can be very helpful.

Younger children need to be given every opportunity to act as the messenger in the school, while children in secondary school may need intensive training about the school, ideally in advance. Parents may need help in developing methods of teaching particularly when visiting hotels and other large public buildings. Some children can be taught to develop a discipline of making up alternative ways of remembering such as mnemonics.

Knowing where things are

Children with profound problems with orientation can have difficulty knowing where they have put things. To the observer it is obvious that the felt tip pen is just on the right hand side but to the child, a pen may be deemed stolen because it is nowhere to be found. Children with this degree of orientational difficulty need to have a dedicated work-station in which the location for the pen is clearly shown. To begin with, a place mat with the patterns of the cutlery can help at meal times. With a lot of hard work these difficulties can be overcome and it is then only when the child is rushed or stressed that the

problems re-emerge.

Visual fatigue

There are many jobs that entail detailed inspection. For example, looking down microscopes, reading X-rays and quality control. It is well recognised that because these tasks involve a lot of mental effort, workers become tired and inaccuracies creep in. Regular well-earned breaks are therefore scheduled to enhance performance. Similarly, children with visual impairment due to damage to the brain become fatigued, and their performance drops off when they have worked hard. The visual system can be fatigued, especially in children with limited vision. There can be periods of remarkable lucidity when the visual system appears to work well, interspersed with periods when the child does not seem to see at all well.

On a lesser scale children get tired much more easily if what they are being shown is a struggle to see, because it is at the limit of their perception, for any of the reasons already described. For all children who fatigue quickly, the first thing to do is to simplify the visual information by enlargement and by removing clutter and pattern in the child's working vicinity. This can often give gratifying results, which can often be enhanced by ensuring that all forms of communication are clear and paced at the speed at which attention is maintained. By diminishing the amount of information a child has to handle,

both in space and in time, and ensuring that everyone working with the child is aware of the limits of detail, complexity and speed of communication that the child can cope with, the number and duration of periods during which the child is inattentive can diminish.

Visual Memory

Our ability to remember what we have seen is very important. The initial part of the process is carried out by the inner parts of the temporal lobes of the brain. If visual memory is impaired due to damage to the temporal lobes and adjacent areas where visual memories are formed, it is not surprising that such tasks as copying are difficult. One strategy worth considering is to encourage the child with a poor visual memory to speak out loud (initially) and then to speak with imagined speech, about what has been seen, so that auditory memories are recruited as well. This can in turn help such activities as copying down information.

Thinking in threes – an approach to mastering the portfolio

A large proportion of the brain is devoted to vision. This means that when there is damage to the brain, vision is commonly affected. The severity of visual impairment can range from being profound, to disorders of visual perception only, with a wide range of combinations of visual difficulties in between. The subject is complex, but it is important to ensure that the topic is fully addressed

when the educational assessment team assesses an affected child and plans management. This requires a mental checklist. Thinking in threes, provides a practical way of doing so (Table 1).

The visual system is made up of *three principal components*: Primary visual processing, the dorsal, and the ventral streams. *For each component there are three principal aspects of vision*, each of which can be impaired in any combination and to any degree. These nine principal visual functions serve *three principal visual needs*, guidance of movement of the body, ability to access information, and the facility to interact socially.

This approach naturally provides 27 concept frameworks that can easily be brought to mind when addressing the *three approaches to (re-)habilitation* namely:

Compensation for the visual difficulties for example, enlarging or magnifying educational material for low visual acuity, or reducing the number of elements to be seen to cater for dorsal stream dysfunction.

Substitution includes the use of tactile approaches such as Braille for material that cannot be seen due to low vision, or recognised due to object and shape agnosia.

Restitution entails training to restore lost functions and is instituted when, for example, seizures, or a blocked shunt have led deterioration of vision that can potentially be restored by appropriate treatment.

Finally, there are three additional elements to add into the equation when considering habilitation and these are impairments of contrast sensitivity, visual memory, and visual imagination.

Table 2 lists some of the difficulties due to cerebral visual impairment along with approaches that can be used to help.

Conclusion

Visual impairment due to damage to the brain is common in children. Affected children can be identified and managed well by being alert to the possibility that it might be present, and carefully evaluating all those in whom it is suspected.

References

Dutton, G.N., Saeed, A., Fahad, B., Fraser, R., McDaid, G., McDade J, Mackintosh, A., Rane, T, Spowart, K. (2004). Association of binocular lower visual field impairment, impaired simultaneous perception, disordered visually guided motion and inaccurate saccades in children with cerebral visual dysfunction-a retrospective observational study. *Eye (Lond)*, 18, 27-34.

Dutton, G.N. & Bax, M. (2010). Visual impairment in children due to damage to the brain. *Clinics in Developmental Medicine No 186*. MacKeith Press, London.

Fazzi E., Signorini S.G., Bova, S.M., La Piana, R., Ondeï, P., Bertone, C., Misefari, W., Bianchi, P.E., (2007). Spectrum of visual disorders in children with cerebral visual impairment. *Journal of Child Neurology*, 22, 294-301.

Good W.V., Hou C., Norcia A.M., (2012). Spatial contrast sensitivity vision loss in children with cortical visual impairment. *Investigative Ophthalmology and Visual Science*, 53, 7730-7734.

Goodale. M., Milner, D. (2004). *Sight Unseen: an exploration of conscious and unconscious vision*. Oxford University Press, Oxford

Guzzetta. A., Tinelli, F., Del Viva, M.M., Bancale, A., Arrighi, R., Pascale, R.R., Cioni, G.,(2009). Motion perception in preterm children: role of prematurity and brain damage. *Neuroreport*, 20, 1339-1343.

Milner, D., Goodale, M. (2006). *The visual brain in action* (second edition) Oxford University Press. Oxford

O'Hare, A.E., Dutton. G.N., Green. D., Coull. R. (1998). Evolution of a form of pure alexia without agraphia, in a child sustaining occipital lobe infarction at 2 1/2 years.

Developmental Medicine and Child Neurology, 40, 417-420.

Ortibus, E.L., De Cock, P.P., Lagae, L.G. (2011). Visual perception in preterm children: what are we currently measuring? *Pediatric Neurology*, 45, 1-10.

Pavlova, M., Staudt, M., Sokolov. A., Birbaumer, N., Krägeloh-Mann, I. (2003). Perception and production of biological movement in patients with early periventricular brain lesions. *Brain*, 126, 692-701.

Saunders, K.J., McClelland, J.F., Richardson, P.M., Stevenson, M. (2008). Clinical judgement of near pupil responses provides a useful indicator of focusing ability in children with cerebral palsy. *Developmental Medicine and Child Neurology*,

50, 33-37.

Saunders, K.J., Little, J.A., McClelland, J.F., Jackson, J. (2010). Profile of refractive errors in cerebral palsy: impact of severity of motor impairment (GMFCS) and CP subtype on refractive outcome. *Investigative Ophthalmology and Visual Science*, 51, 2885-2890.

Section 3 Tables

Table 1 Topics, grouped in threes, to hold in mind when assessing and planning management of a child suspected to have CVI.

1. Visual system

A. Primary vision

- | | | |
|-----------|-------------------|----------------------|
| I. Acuity | II. Visual fields | III. Seeing movement |
|-----------|-------------------|----------------------|

B. Dorsal stream

- | | | |
|--------------------|-------------------|-----------------------|
| I. Visual guidance | II. Visual search | III. Visual attention |
|--------------------|-------------------|-----------------------|

C. Ventral stream

- | | | |
|---------------------|-----------------------|------------------|
| I. Face recognition | II. Shape recognition | III. Orientation |
|---------------------|-----------------------|------------------|

2. Principal Visual functions

- | | |
|---------------------------|----------------|
| I. Guidance of movement | arms / legs |
| II. Access to information | near / distant |
| III. Social interaction | near / distant |

3. (Re)-Habilitation

- I. Compensation
- II. Substitution
- III. Restitution (of functions lost following complications eg a blocked shunt)

4. Additional elements

- I. Contrast sensitivity perception.
- II. Visual memory
- III. Visual imagination

Table 2 Table outlining the visual difficulties that children with cerebral palsy can have and approaches which can help them. (Adapted from [2]: McKillop E, Dutton GN. Impairment of vision in children due to damage to the brain: a practical approach. British and Irish Orthoptic Journal. 2008; **5**: 8-14.)

Problem	Approaches
Reduced clarity of vision.	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
Lack of vision or attention on one side	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

	<p>Careful guidance around new environments</p> <p>Training in crossing roads</p> <p>Turn plate to eat food</p>
Lack of vision down below	<p>As with hemianopia</p> <p>Regularly looking down to check the ground ahead</p> <p>Tactile guide to ground height</p>
Impaired ability to move the eyes	<p>Movement of the head</p> <p>Enlarging text</p> <p>Double spacing text</p> <p>Tracing of text with a finger or ruler</p>
Impaired ability to see movement	<p>Television programmes with limited movement</p> <p>Educational material with limited movement</p> <p>Careful training or guidance in crossing roads.</p>
Difficulty finding a toy in a toy box or an item of clothing in a pile or wardrobe	<p>Separate storage of favourite items</p> <p>Organised storage systems</p> <p>Always store in same location</p> <p>Avoid clutter</p> <p>Colour coding and labels</p>
Difficulty finding an object on a patterned	<p>Use plain carpets, bedspreads and decoration.</p>

background.	
Difficulty finding food on a plate	Avoid patterned plates Avoid sauces/ gravy Separate food portions
Problems seeing a distant object	Use zoom on video/ digital camera to view
Problems reading	Enlarge text Double space text Masking surrounding text Computer programs to present information
Difficulty finding someone in a group.	Wear obvious identifier Always stand in same location Waving 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 Bannister Mark edge of stairs Good lighting Tactile guides to gage the height of the ground

	<p>Approach obstacles with "Look- Slow- Check- Go"</p> <p>Activities to improve coordination</p>
Inaccurate visually guided reach	<p>Reaching beyond an object to gather it.</p> <p>Activities to improve coordination</p> <p>Occupational therapy</p>
Difficulty 'seeing' when talking at the same time	<p>Limit conversation when walking</p> <p>Identify obstacles by tactile stimulation</p>
Frustration at being distracted	<p>Limit distraction</p> <p>Minimise background clutter</p> <p>Minimise background activity.</p> <p>Quiet table at school</p>
Difficulty recognising people and photographs	<p>Introductions</p> <p>Training in identifying voices</p> <p>Consistent identifiers worn</p> <p>Training to recognise identifiers</p>
Difficulty recognising shapes and objects	<p>Training to identify and recognise identifiers. Training in tactile recognition</p>
Difficulty reading facial expression	<p>Training in recognising facial expressions</p> <p>Expression of mood by tone of voice</p> <p>Explanation of mood in words</p>

Getting lost in known places	Training in orientation. Encouraging leading Incorporating landmarks in Mnemonics /Poems
Difficulty in new environments	Training in orientation. Encourage exploration -Visit at quiet times -Hide and Seek -Treasure Hunts
Visual fatigue Prolonged visual processing	Minimise clutter Reduce distractions Reduce detail and complexity Well earned breaks
Social problems	Good understanding and support at school. Identify problems and solutions Encourage child to overcome them Well known informed peer group Find activities child enjoys and can excel in