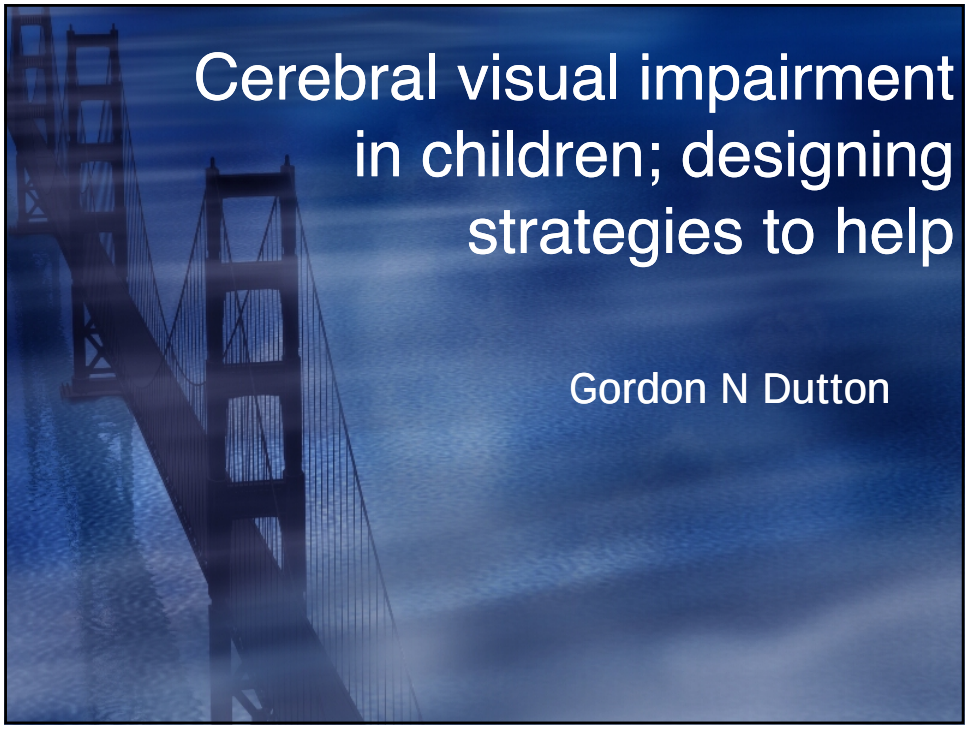


Background

- Set up pediatric neuro-ophthalmology clinic in 1988
- Increasing number of referrals of older children with profound but undiagnosed visual perceptual problems
- In 1990 Vision assessment team was set up
- By 1995 clear pattern was emerging of a range of patterns of visual difficulty*

*Dutton G, Ballantyne J, Boyd G, Bradnam M, Day R, McCulloch D, Mackie R, Phillips S, Saunders K. Cortical visual dysfunction in children: a clinical study. Eye 1996, **10:** 302-309.

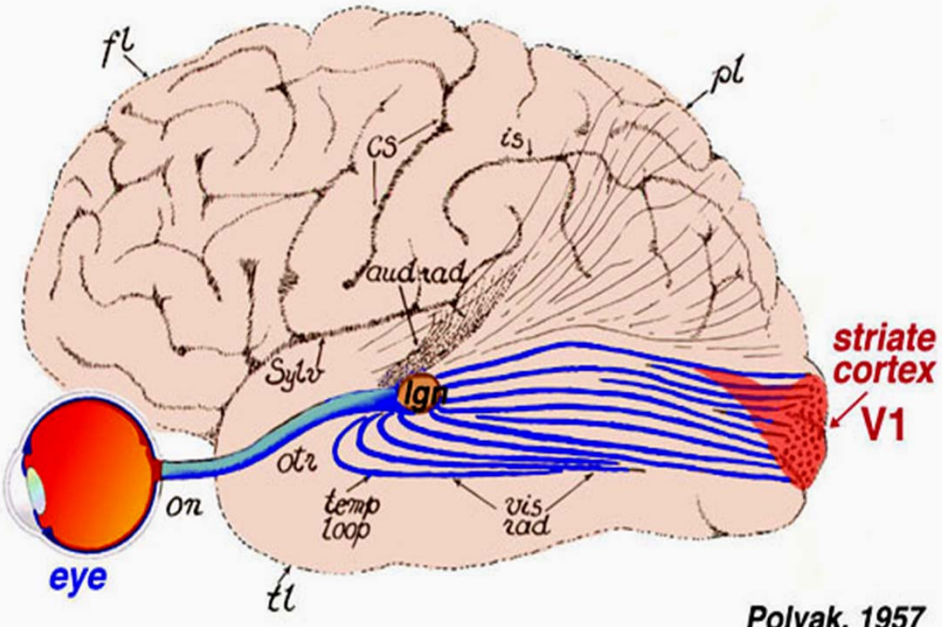
Background

- At the same time Lena Jacobson (Stockholm)* described the same symptom complex in children with PVL (often with nystagmus)
- And Milner and Goodale** published their seminal treatise on the dorsal and the ventral streams mediating higher visual function
- Since then a number of studies corroborated these findings

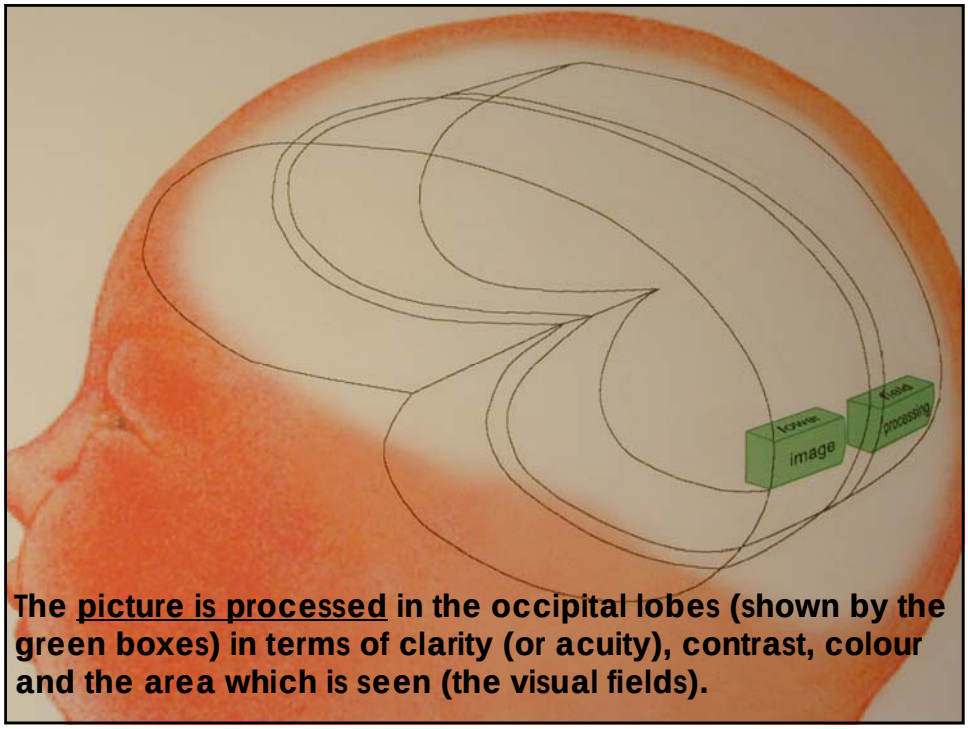
*For review see: Jacobson LK, Dutton GN. Periventricular leukomalacia: an important cause of visual and ocular motility dysfunction in children. *Surv Ophthalmol* 2000; **45:** 1-13.

Milner AD, Goodale MA (2006) The Visual Brain in Action. 2nd Edition. Oxford University Press, Oxford (1st edition in 1995**)

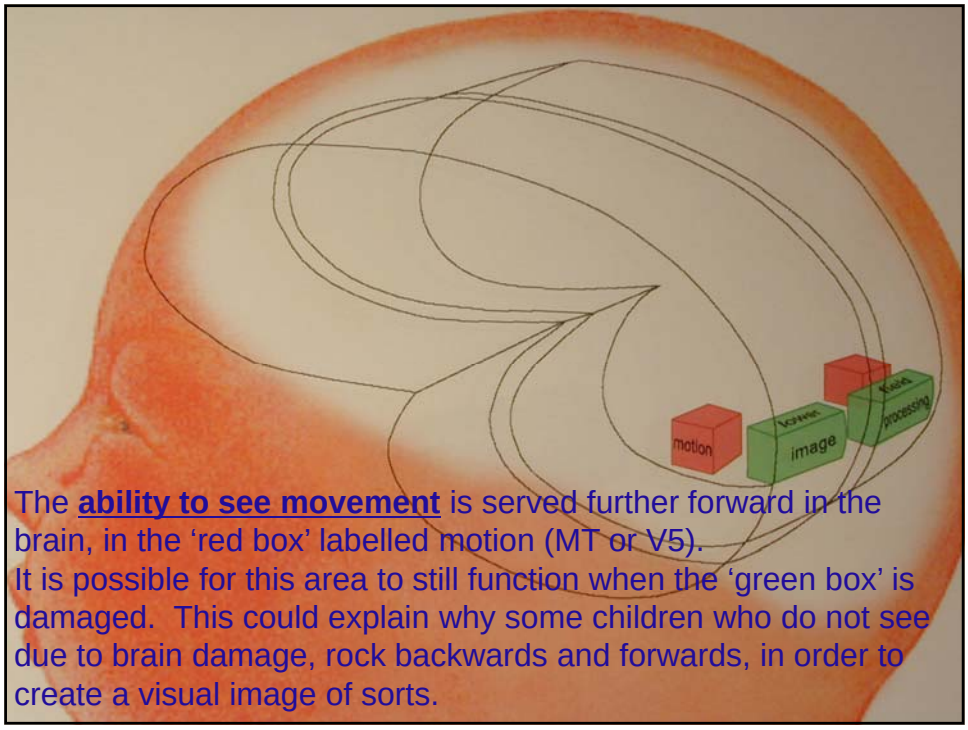
Goodale MA, Milner AD. (2004) Sight Unseen: An Exploration of Conscious and Unconscious Vision. Oxford: Oxford University Press.



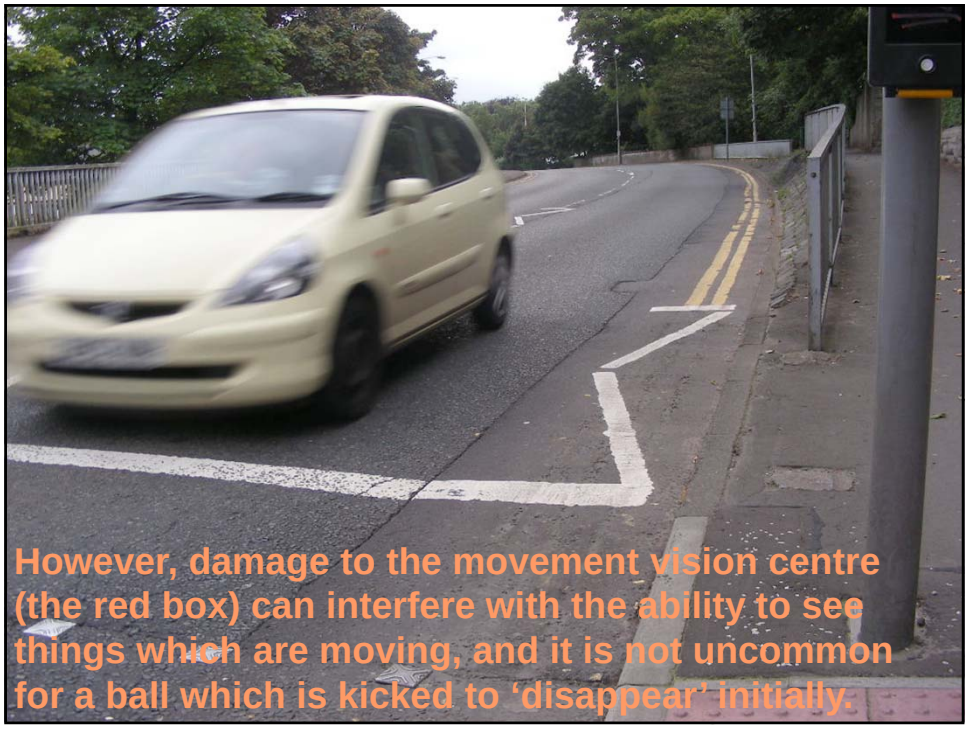
The model for cortical visual impairment -



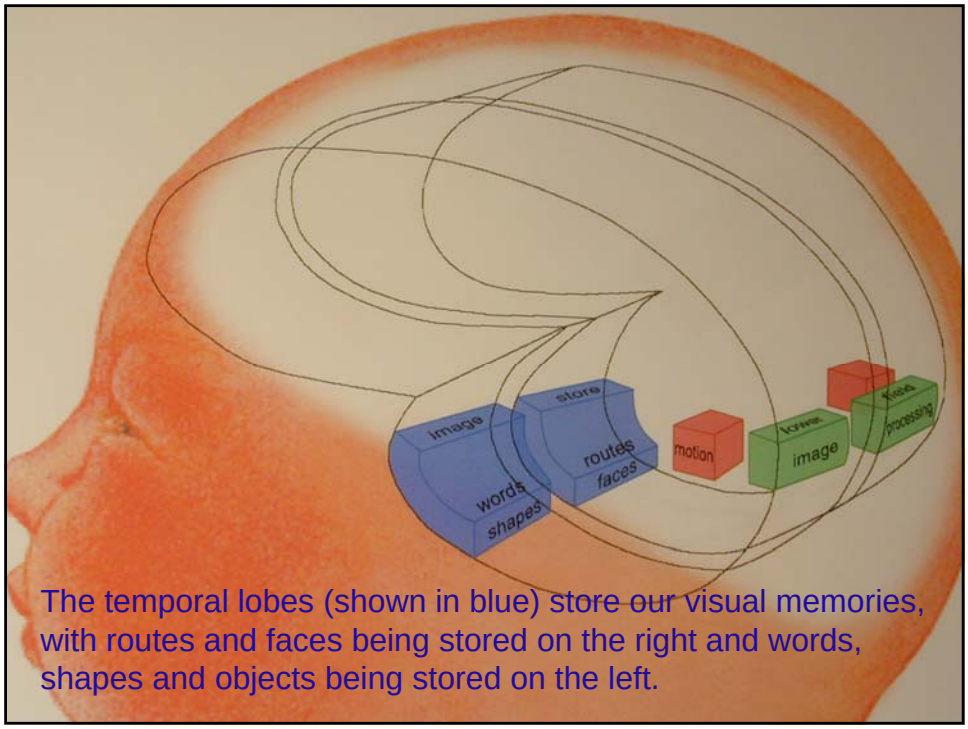
The <u>picture is processed</u> in the occipital lobes (shown by the green boxes) in terms of clarity (or acuity), contrast, colour and the area which is seen (the visual fields).



The **<u>ability to see movement</u>** is served further forward in the brain, in the 'red box' labelled motion (MT or V5).
It is possible for this area to still function when the 'green box' is damaged. This could explain why some children who do not see due to brain damage, rock backwards and forwards, in order to create a visual image of sorts.

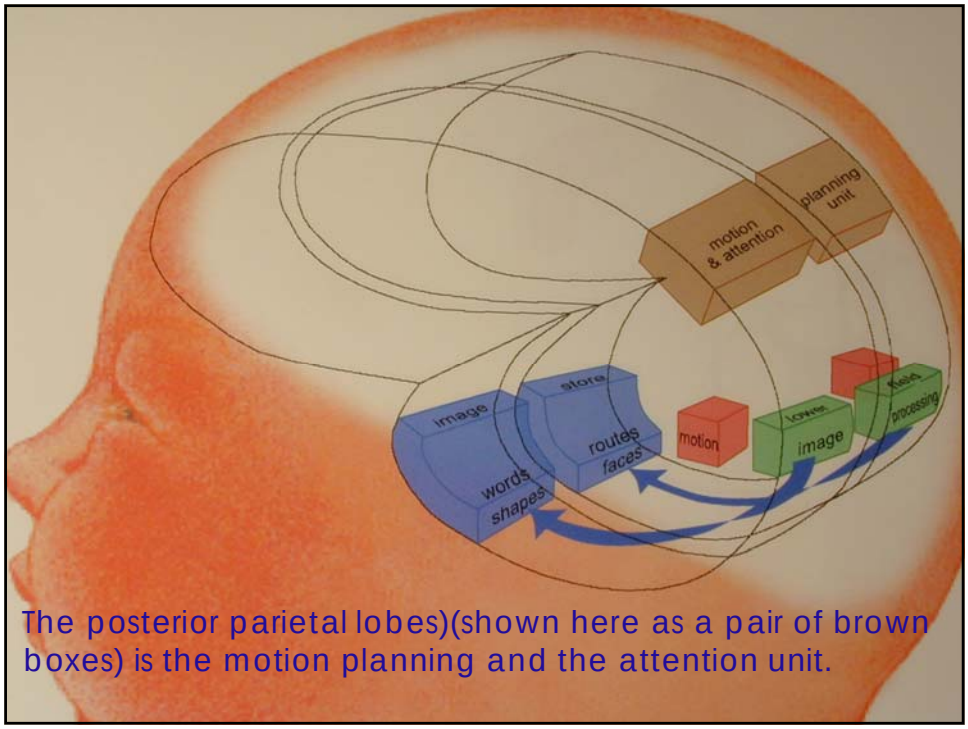


However, damage to the movement vision centre (the red box) can interfere with the ability to see things which are moving, and it is not uncommon for a ball which is kicked to 'disappear' initially.



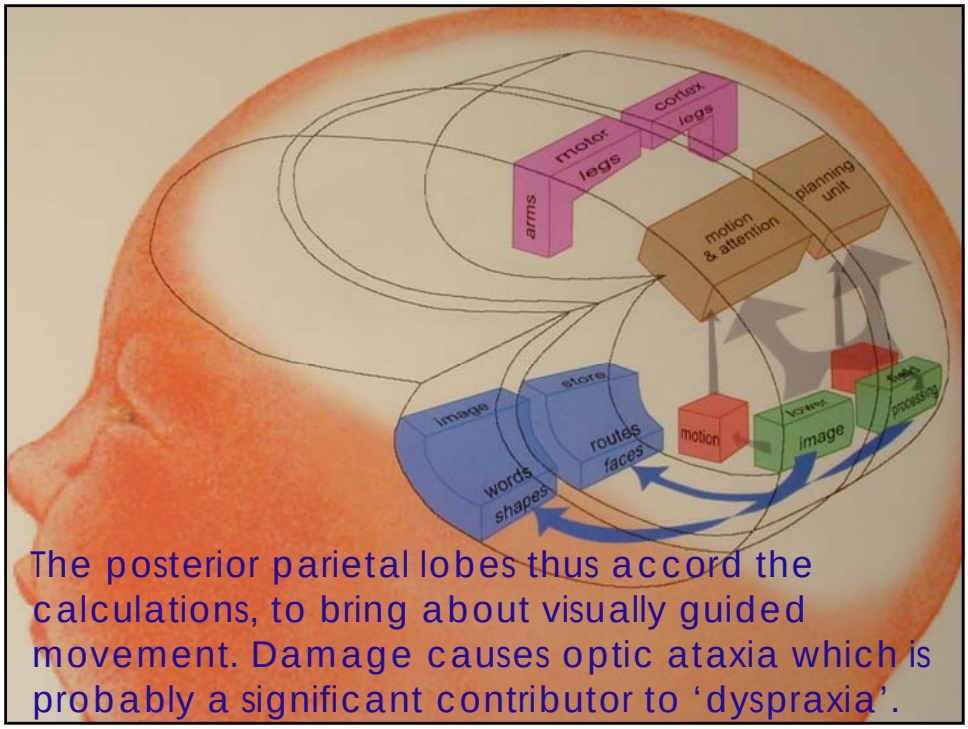
The temporal lobes (shown in blue) store our visual memories, with routes and faces being stored on the right and words, shapes and objects being stored on the left.

So that damage on both sides would interfere with the recognition of both the words. and the person shown in this picture

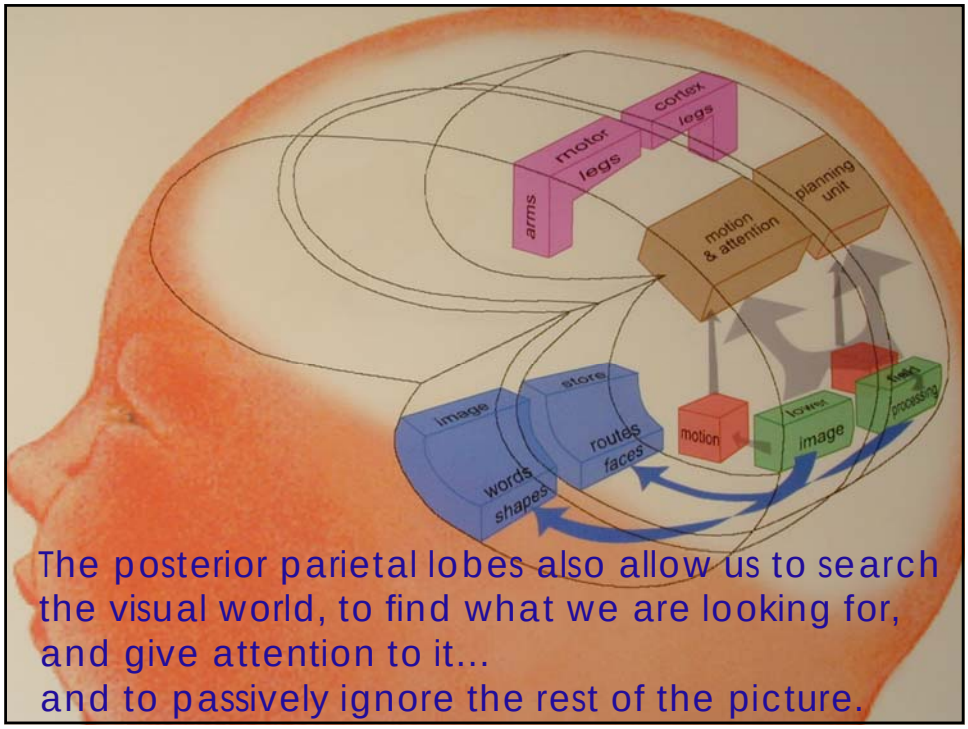


The posterior parietal lobes)(shown here as a pair of brown boxes) is the motion planning and the attention unit.

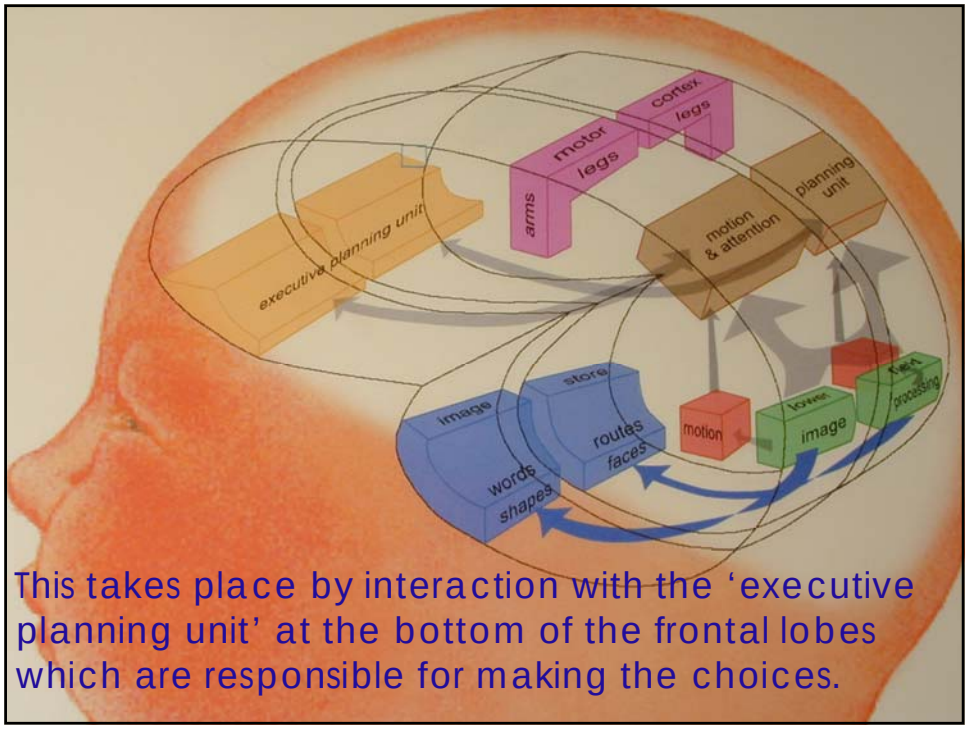
Visual attention and guidance of movement



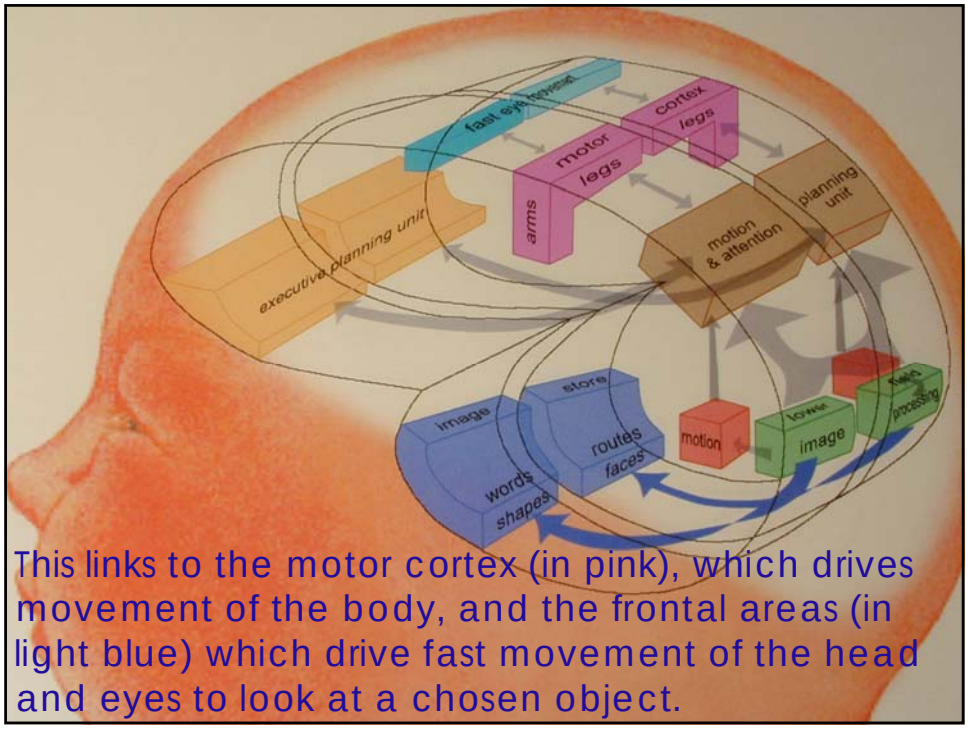
The posterior parietal lobes thus accord the calculations, to bring about visually guided movement. Damage causes optic ataxia which is probably a significant contributor to 'dyspraxia'.



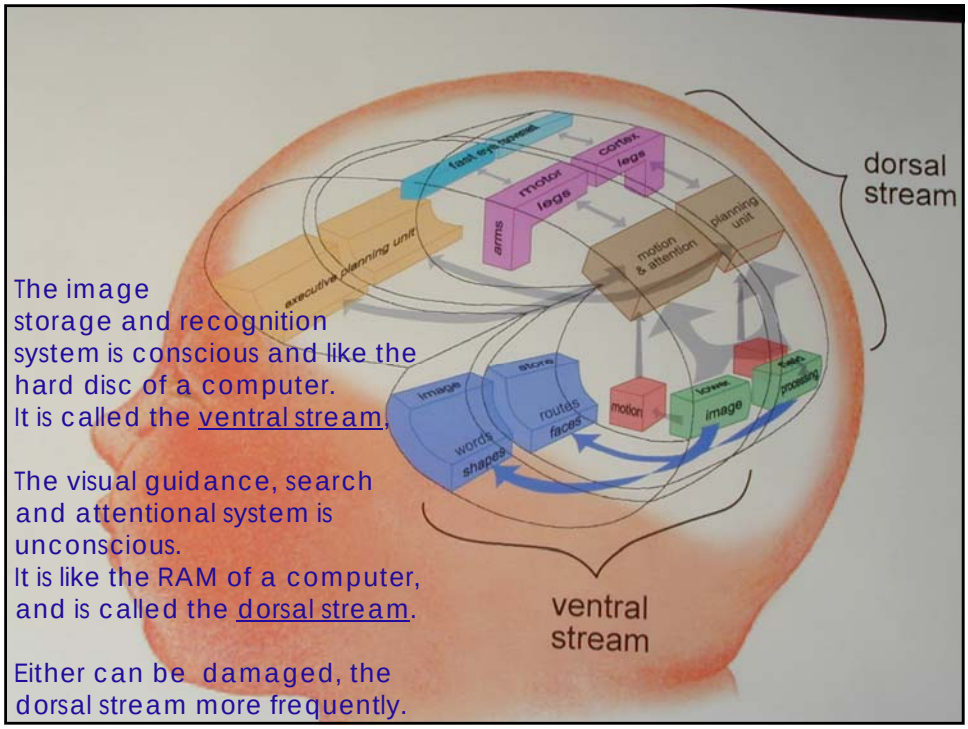
The posterior parietal lobes also allow us to search the visual world, to find what we are looking for, and give attention to it...
and to passively ignore the rest of the picture.



This takes place by interaction with the 'executive planning unit' at the bottom of the frontal lobes which are responsible for making the choices.



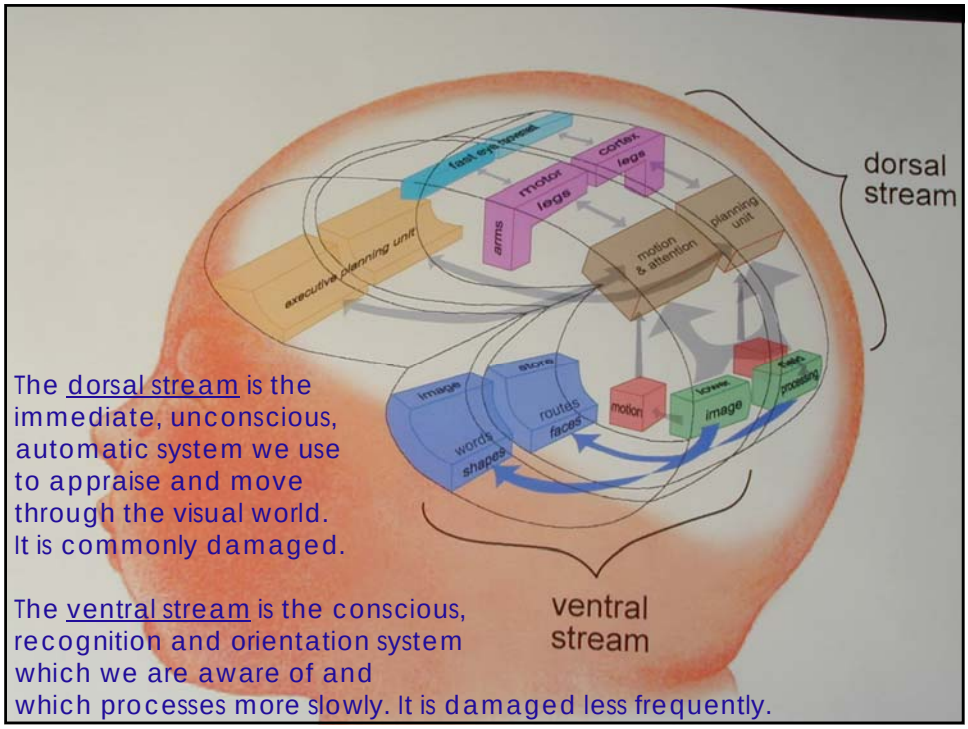
This links to the motor cortex (in pink), which drives movement of the body, and the frontal areas (in light blue) which drive fast movement of the head and eyes to look at a chosen object.



The image storage and recognition system is conscious and like the hard disc of a computer. It is called the ventral stream.

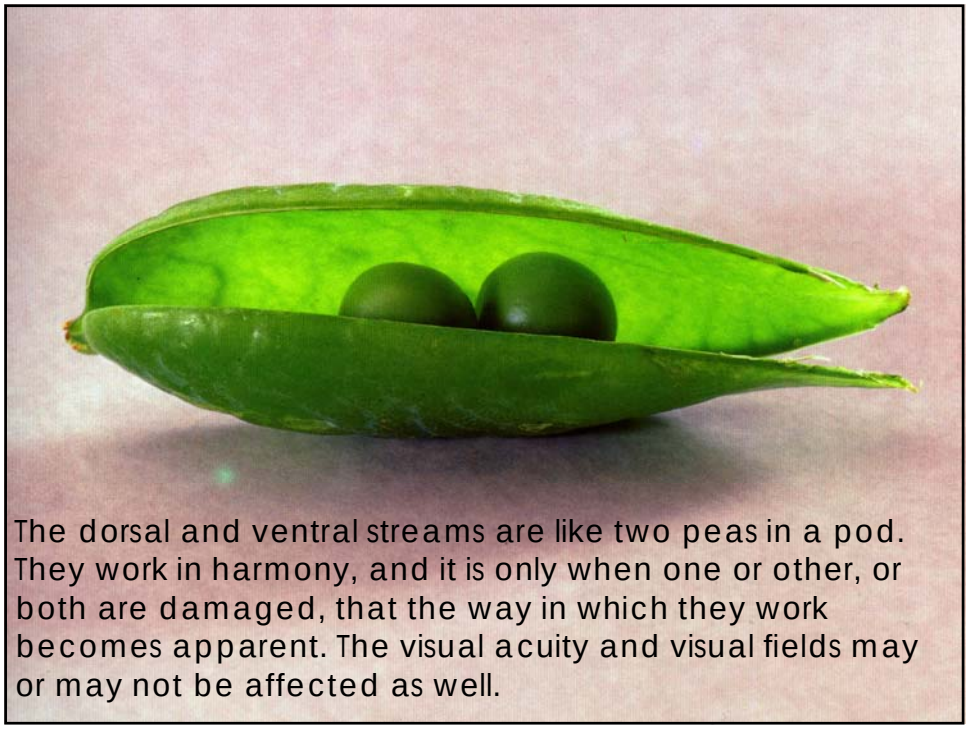
The visual guidance, search and attentional system is unconscious. It is like the RAM of a computer, and is called the dorsal stream.

Either can be damaged, the dorsal stream more frequently.



The dorsal stream is the immediate, unconscious, automatic system we use to appraise and move through the visual world. It is commonly damaged.

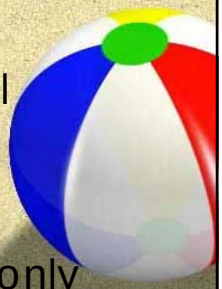
The ventral stream is the conscious, recognition and orientation system which we are aware of and which processes more slowly. It is damaged less frequently.



The dorsal and ventral streams are like two peas in a pod. They work in harmony, and it is only when one or other, or both are damaged, that the way in which they work becomes apparent. The visual acuity and visual fields may or may not be affected as well.

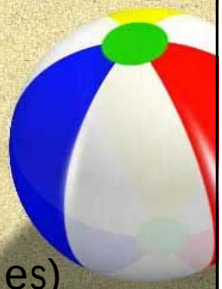
Children with cerebral visual impairment include:

- Those with profound visual impairment
- Those with and without cerebral palsy
- Those with poor acuities +/- visual field impairment
- Those with poor acuities +/- visual field impairment and perceptual problems
- Those with perceptual problems only



Profound 'cortical' visual impairment and blindsight :

- Usually have 4 limb cerebral palsy
- But may be mobile with 'travel vision'
- Fatiguable 'low level' blindsight can be used to aid feeding
- It may ultimately develop into form vision as a form of late DVM
- It may be accompanied by affective blindsight (smiling at smiles)

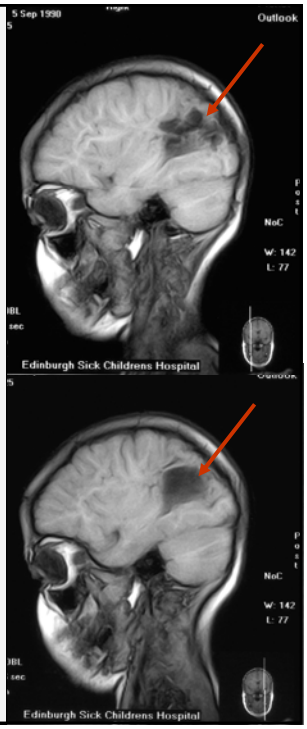


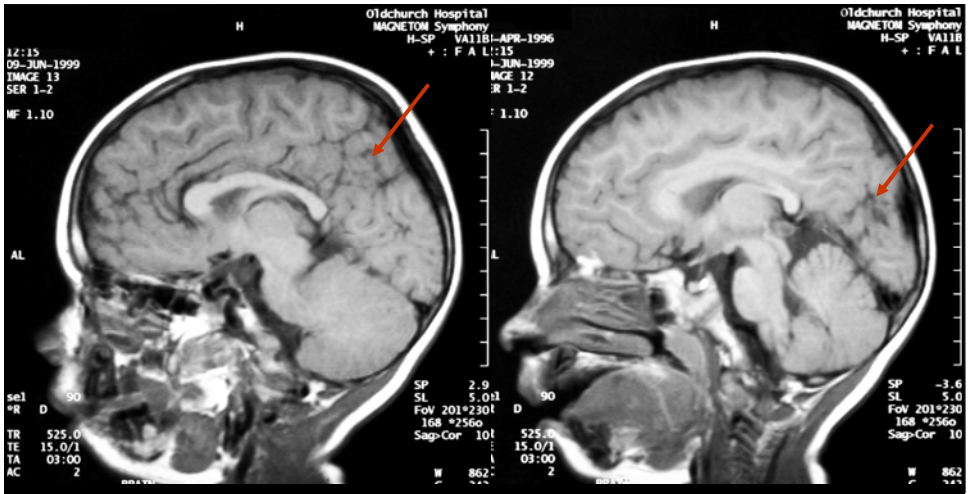
These scans are of two children who have:

Slightly reduced acuities.
Absent lower visual fields
Impaired visual guidance of movement.
Ability to read, only when adjacent text is covered.
Inability to find someone in a group, or a toy in a toy box, or an item of clothes in a pile.

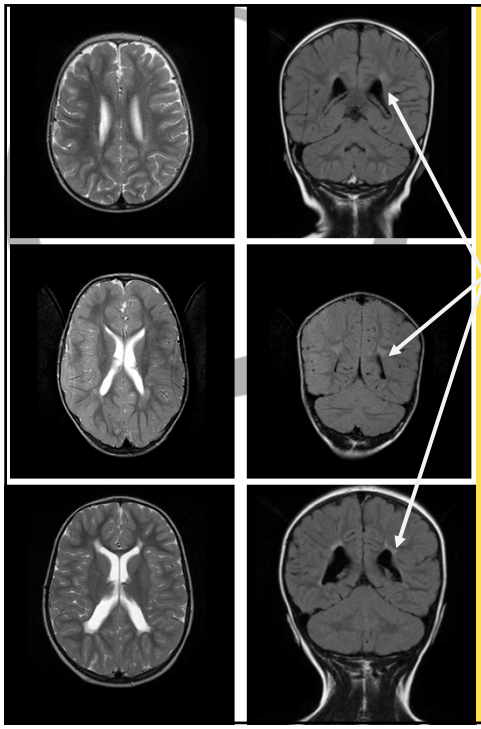
They are in main stream school.

They have posterior parietal / dorsal stream damage, with features of Balint's Syndrome.





These are scans of another child, who has exactly the same problems with damage in the same area.
The diagnosis of dorsal stream damage had been missed for a long time in all three of these children. (Two of these cases have been published.)



These pairs of MRI scans* are of three children who have very similar sets of perceptual visual difficulties, with near normal visual acuities.

Remarkably, they have focal pathology in the white matter in the posterior parietal area.

This is the commonest cause of poor vision in children now seen in Glasgow. (Now that we know to look for it!)

*Saidkasimova et al. Cognitive visual impairment with good visual acuity in children with posterior periventricular white matter injury: A series of 7 cases. J AAPOS. 2007; 11: 426-30.

Correction of even small amounts of long sightedness can benefit these children considerably, because these children not only have lack of accommodation to compensate for their longsightedness but they also benefit from the reduction of crowding provided by the spectacle magnification.

The Visual Limitations Caused by Damage to the Brain in Children Include:

- Deficits in accommodation (>40%) & eye movements
- Those of impaired primary processing of the image which may affect
  - Visual acuity, contrast sensitivity, colour vision, visual fields, and the perception of movement
- Those affecting the dorsal stream, impairing
  - The ability to give attention to, and thus see, the components of a crowded visual scene - and -
  - The use of vision to guide movement
- Those affecting the ventral & ventro-dorsal streams

(Which are less common) Involving:

- Recognition of what is being looked at
- and recognition for route finding

The Visual Limitations Caused by Damage to the Brain in Children Lead to problems:

- Accessing information
  - Whether it is in the distance, such as a letter box or a tree
  - Or whether it is for near, such as pictures and text
- With visual guidance of movement
  - Whether it is the guidance of the upper limbs, to reach out and pick something up
  - Or whether it is the guidance of the lower limbs, to walk freely and confidently
- And with social interaction. It may be difficult to
  - To see someone in a group, owing to visual acuity or visual field loss, or dorsal stream problems
  - To recognise their face, owing to poor acuity, contrast sensitivity, or agnosia
  - To see the language within facial expression (for the same reasons)
  - Or even to see a hand in an absent lower visual field, for a hand shake

Functional visual assessment

How the visual acuity, contrast sensitivity, colour vision and visual fields affect daily life, can be determined by measuring visual function, followed by functional visual assessment.

However, finding out about how dorsal stream and ventral stream visual problems interfere with vision can be carried out, both by structured history taking and by carefully watching and Interpreting the child's visual behaviour.

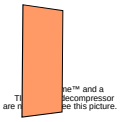
Bumping into door frames, and obstacles like street signs, on one side suggests that visual field impairment or inattention on that side is worth looking for.

Leaving food on one side of the plate and missing traffic from one side,

While missing text on one side and leaving food on the near side of the plate, all suggest such problems.

Rotating the plate to find the food and early training to look to the side for traffic are useful approaches

Modifying the presentation of text on one side and moving the bowl further away, to allow the near side of the plate to be seen are further approaches to deal with such difficulties



QuickTime™ and a
TIFF (LZW) decompressor
are needed to see this picture.

A bright marker on door frames at the child's eye level can help,for lack of attention on one side; and early training in mobility and independence is needed for being out and about.

In the classroom hemianopia or lack of attention on one side may require:

- The teacher to be off centre to the sighted side
- Strategies to ensure that information on the affected side is not missed, which can include:
  - –Using tactile guides on a page
  - –Training in scanning the information
  - –Presenting information displaced to the sighted side

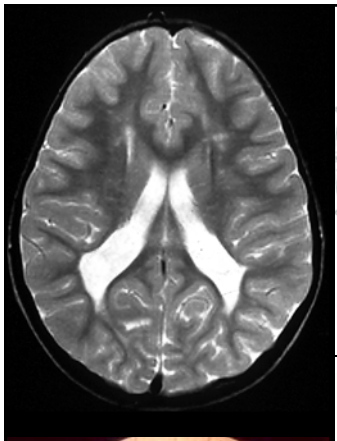
In addition:

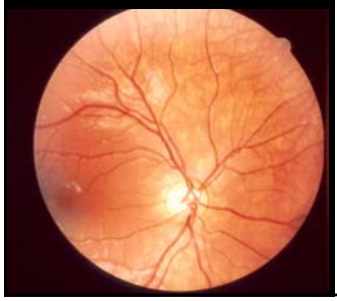
If one cannot see on the right side, the words on the right side of the page may not come into view, so that they are missed. However, when the text is rotated through 90 degrees so that it can be read vertically downwards, the area that is not seen simply covers over the text which has been read so that it does not get in the way.

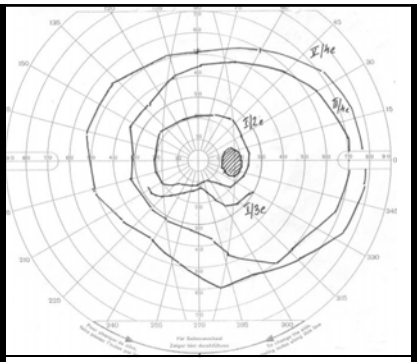
If one cannot see on the left side, the words on the left side of the page disappear as the eye looks to the right side of the page. This makes the next line difficult to find and information can be missed, but when the text is rotated through 90 degrees so it can be read vertically upwards, this no longer happens, as all the text to be read is visible.

QuickTime™ and a
TIFF (LZW) decompressor
are needed to see this picture.

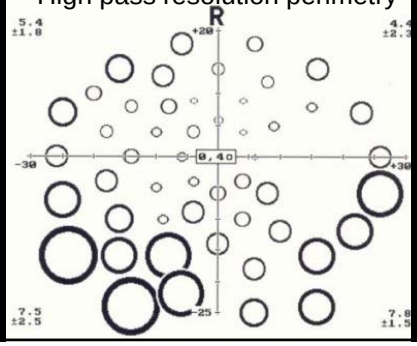
Lower visual field impairment, and impaired visual guidance of movement of the feet, cause problems with steps for which tactile guidance may be needed.







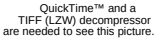
High pass resolution perimetry



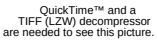
QuickTime™ and a
TIFF (LZW) decompressor
are needed to see this picture.

The floor needs to be kept clear of toys and low obstacles outside may cause problems.

QuickTime™ and a
TIFF (LZW) decompressor
are needed to see this picture.

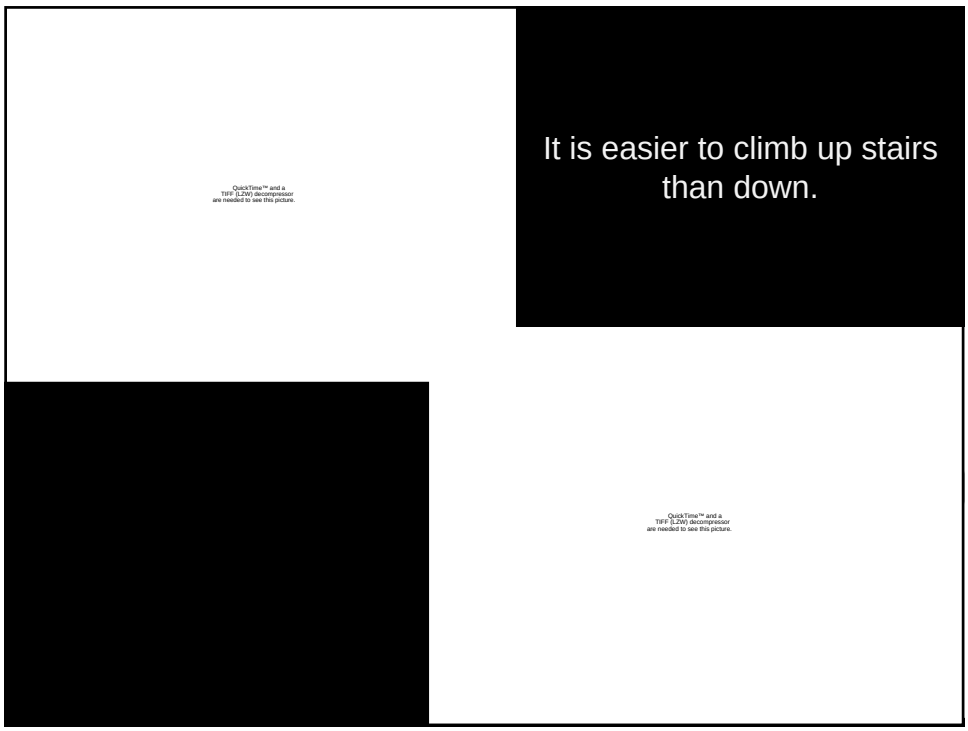


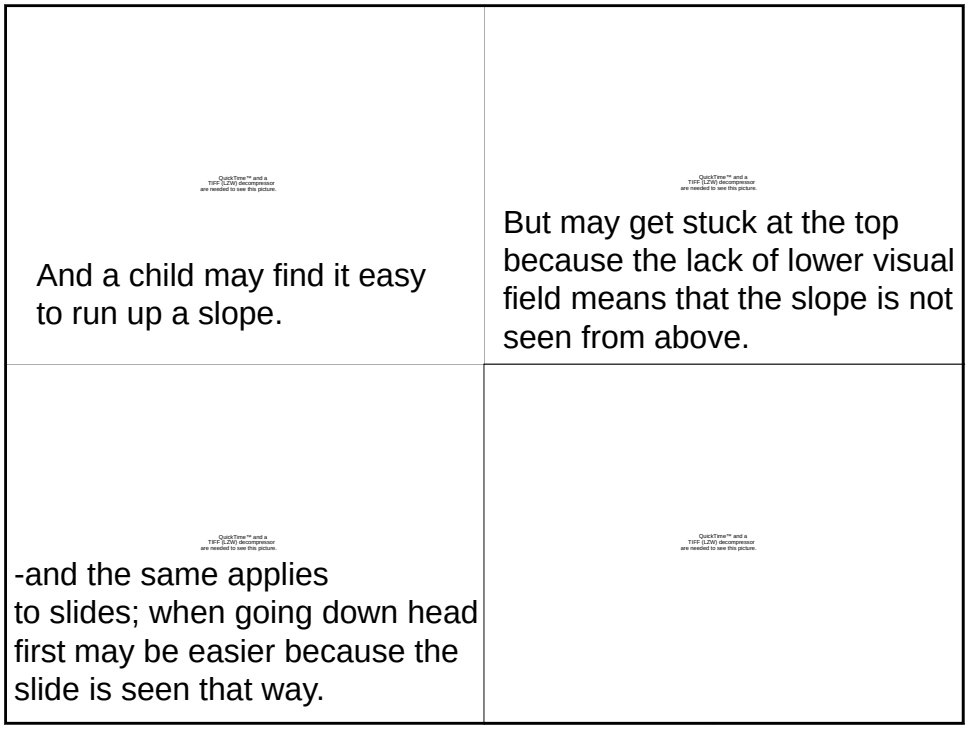
As can be seen from this photograph, the view going down gives less information -



than the view going up, where the side of pavement or step is also visible, which can lead to stepping off the edge of a pavement.

Floor boundaries both inside and out can cause hesitation and a need to explore the boundary with the foot.


It is easier to climb up stairs than down.

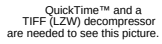

And a child may find it easy to run up a slope.
But may get stuck at the top because the lack of lower visual field means that the slope is not seen from above.
-and the same applies to slides; when going down head first may be easier because the slide is seen that way.

Watching the TV upside down is highly suggestive of lower visual field impairment in a young child!

QuickTime™ and a
decompressor
are needed to see this picture.

QuickTime™ and a
TIFF (LZW) decompressor
are needed to see this picture.

Providing a tactile guide to the height of the ground ahead, whether it is a brick trolley for a younger child - or



- a scooter for an older child gives the child greater independence

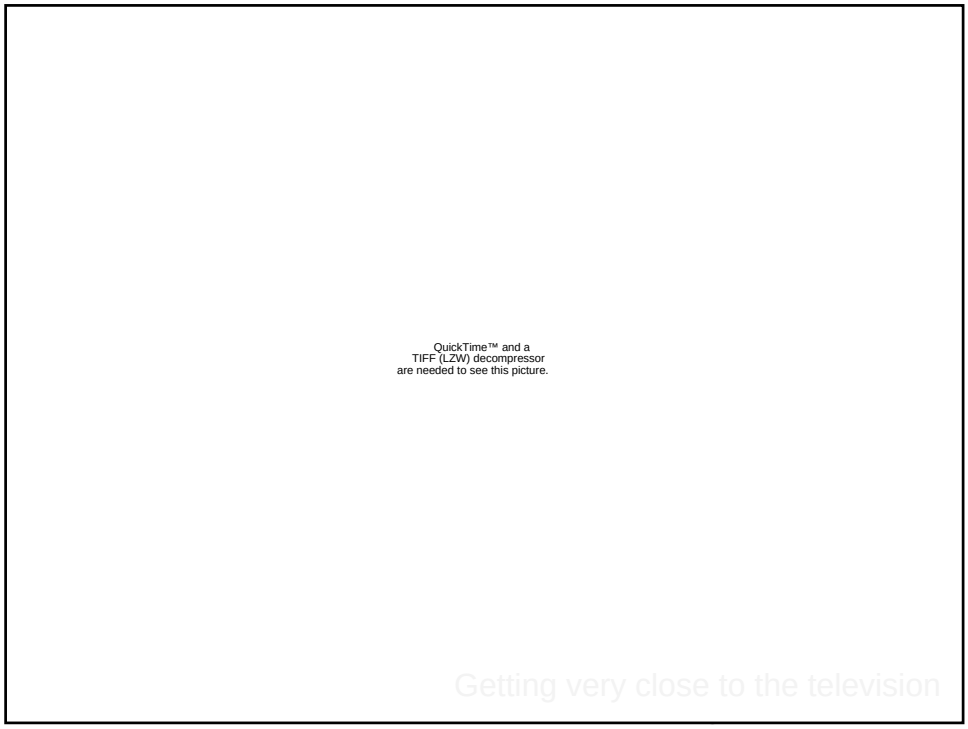
Checking how far the foot has to be extended before it is seen, is a consistent test of the peripheral lower visual field, for the older child.

When walking holding hands with such a child, holding one's arm slightly backwards, like this, gives advance notice of changes in the height of the ground ahead, and keeping the arm straight like this, gives the guidance to the height itself.

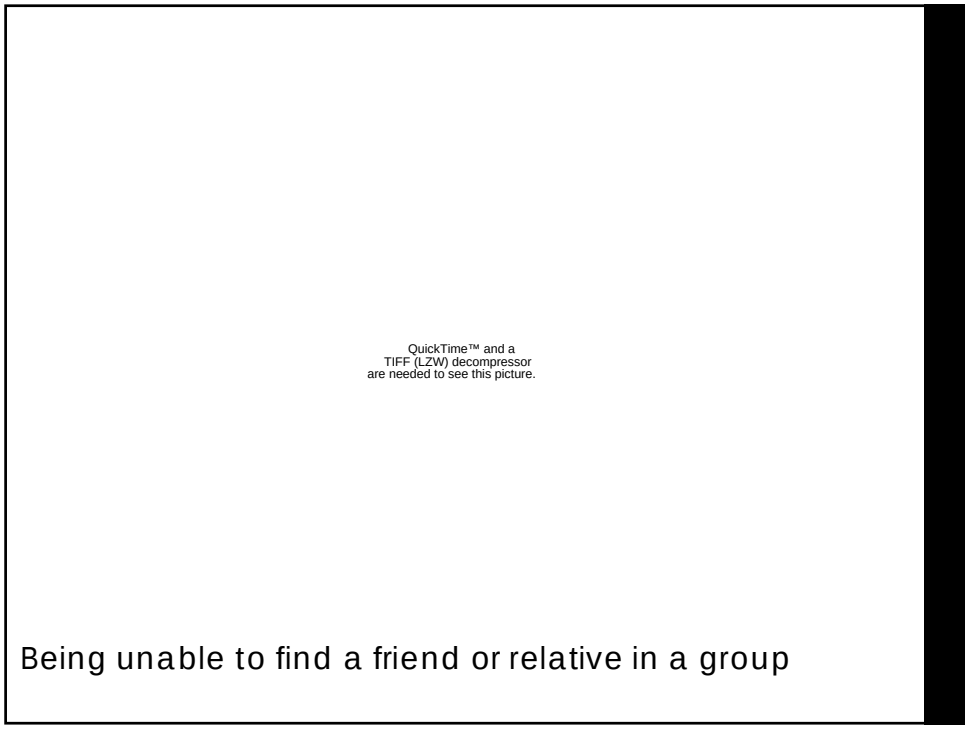
We have also recently realised that lower visual field impairment leads to the child having to lift the foot up high to change shoes, this may be done lying on the back if vision, balance or leg strength are poor.

As is well recognised, a tilted work station, helps to ensure that work at the bottom of the page is not missed, as this decreases the effect of lower visual field impairment.

The features on history taking for dorsal stream dysfunction include:



Getting very close to the television



Being unable to find a friend or relative in a group

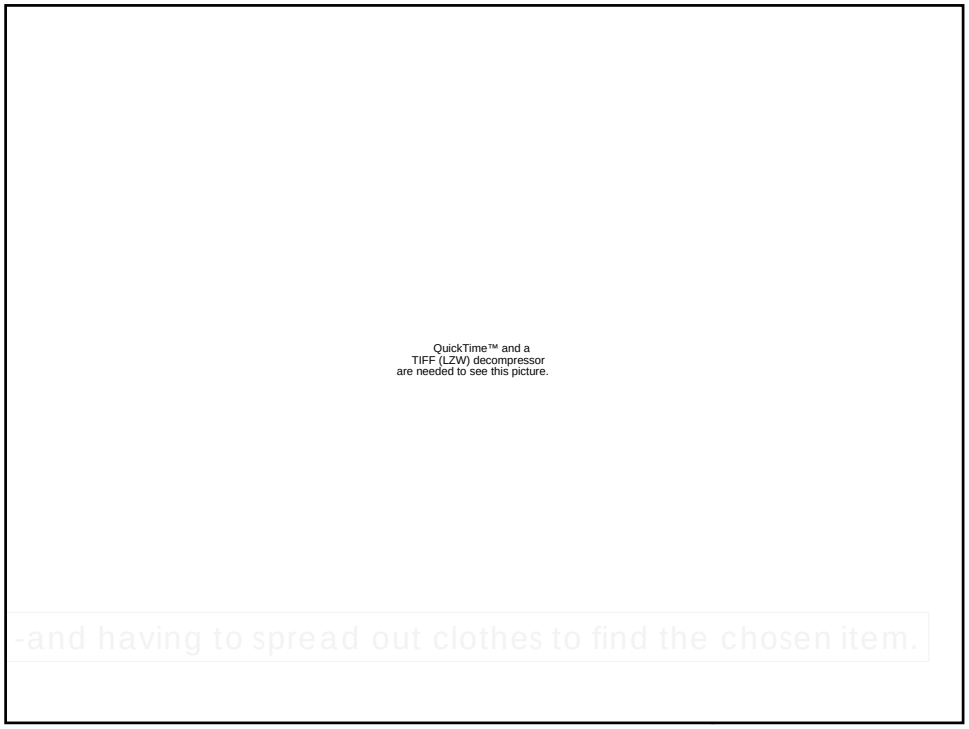
QuickTime™ and a
TIFF (LZW) decompressor
are needed to see this picture.

Having difficulty finding a toy on a patterned background

QuickTime™ and a
TIFF (LZW) decompressor
are needed to see this picture.

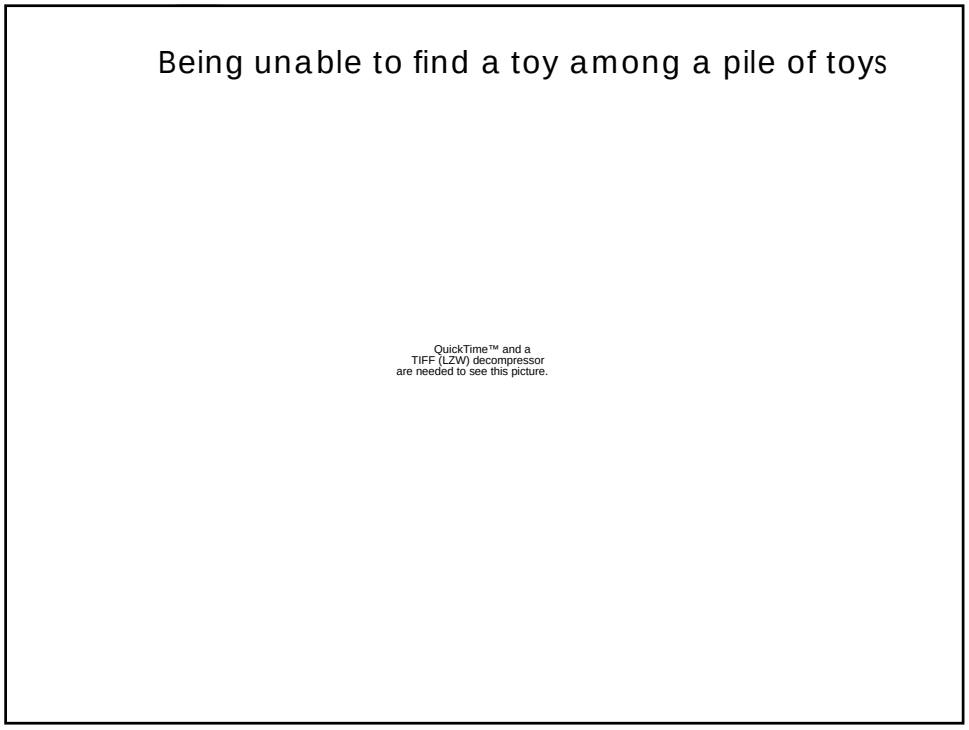
QuickTime™ and a
TIFF (LZW) decompressor
are needed to see this picture.

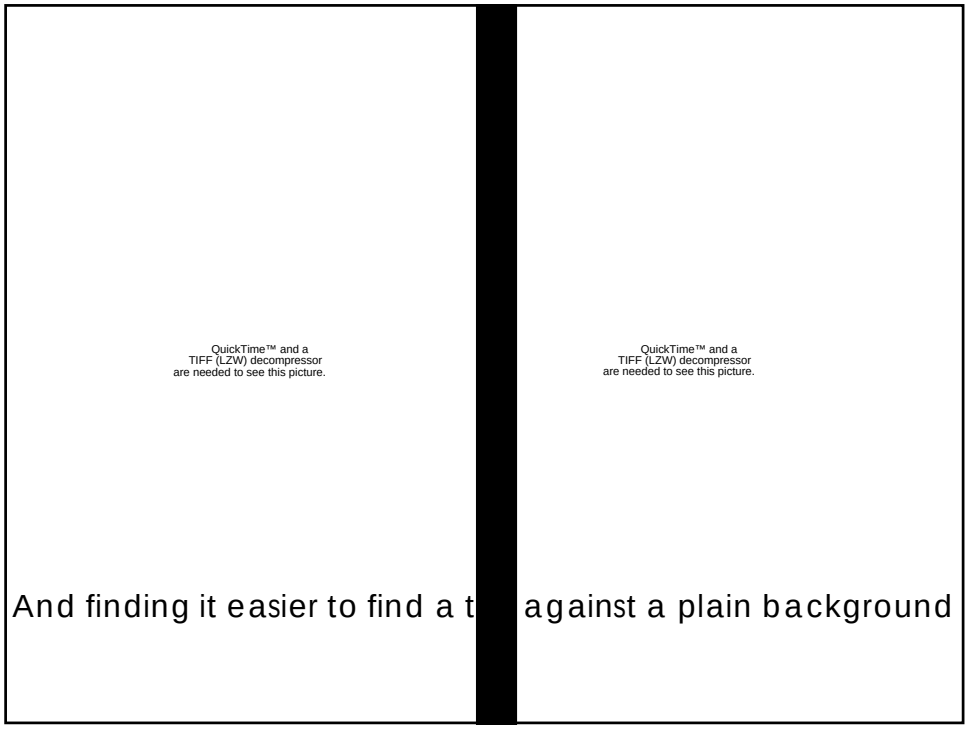
-being unable to find an item of clothing in a pile of clothes

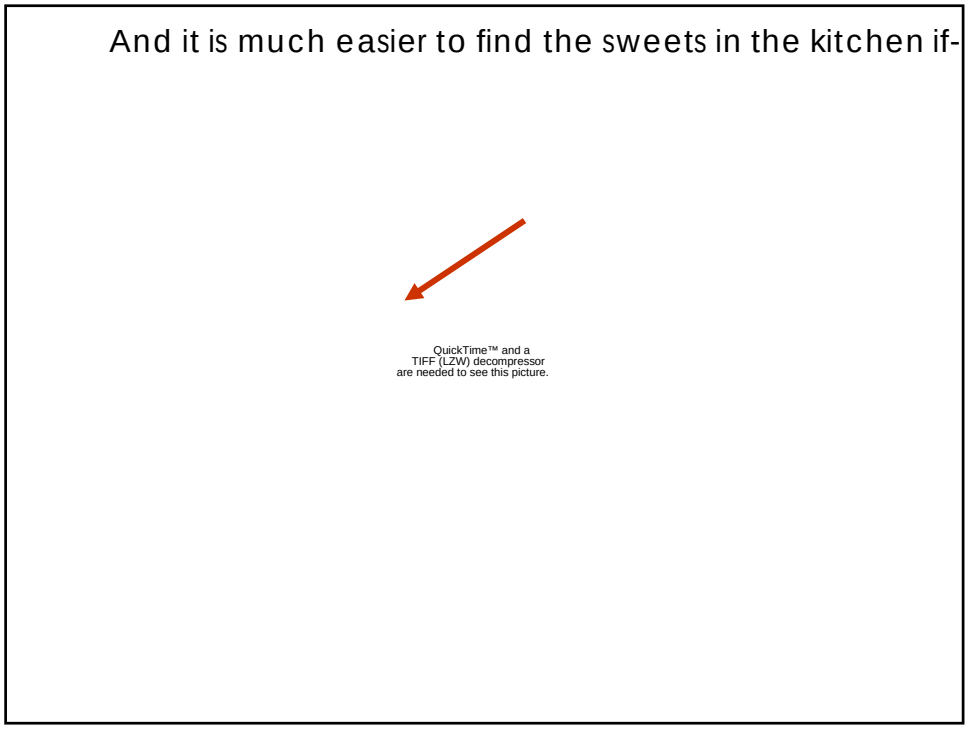


-and having to spread out clothes to find the chosen item.

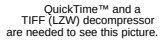
Being unable to find a toy among a pile of toys



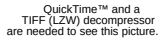

QuickTime™ and a
TIFF (LZW) decompressor
are needed to see this picture.
QuickTime™ and a
TIFF (LZW) decompressor
are needed to see this picture.
And finding it easier to find a t
against a plain background


And it is much easier to find the sweets in the kitchen if-
QuickTime™ and a
TIFF (LZW) decompressor
are needed to see this picture.

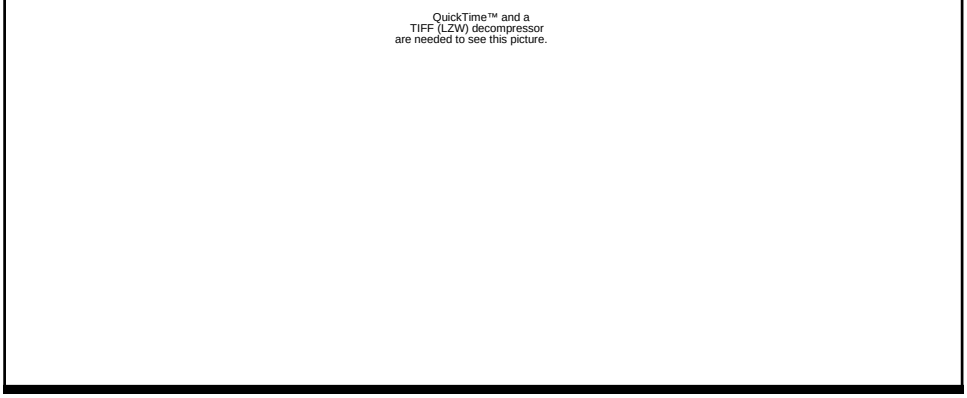
There is no clutter!



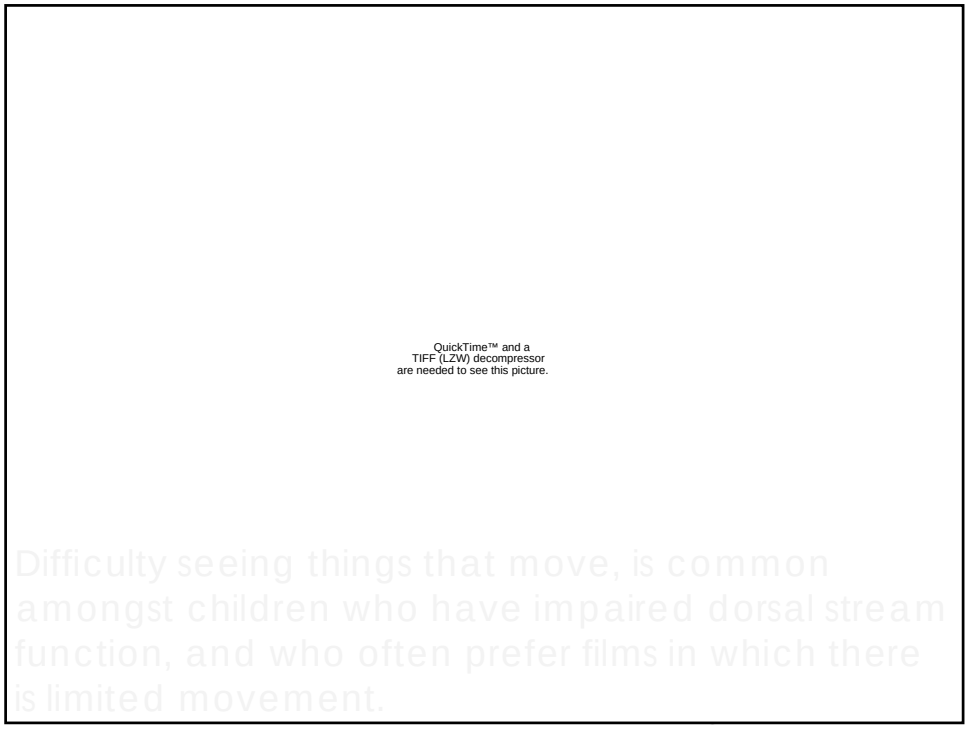
Like this!



The further things are away, the more there is to see (as well their being smaller). Children with dorsal stream dysfunction cannot easily see things which are pointed out in the distance, and can be helped by sharing the screen of a video camera.



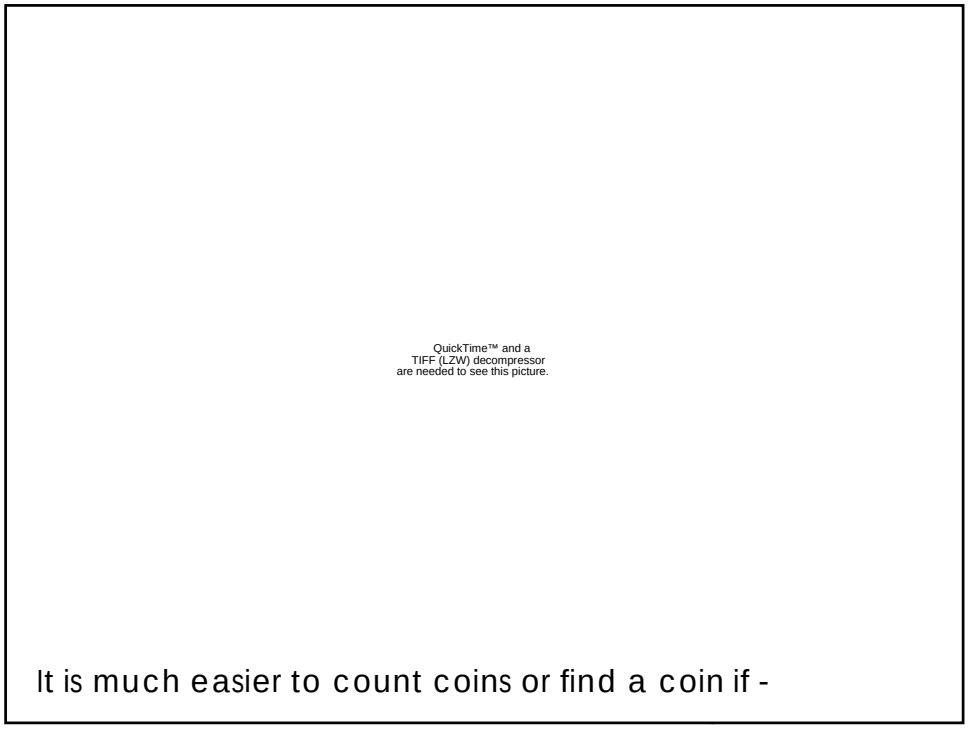
And it is likely to be visual overload, which contributes to difficulty finding the way around, for which it can help by using the horizon, where there is least clutter, to search for landmarks .



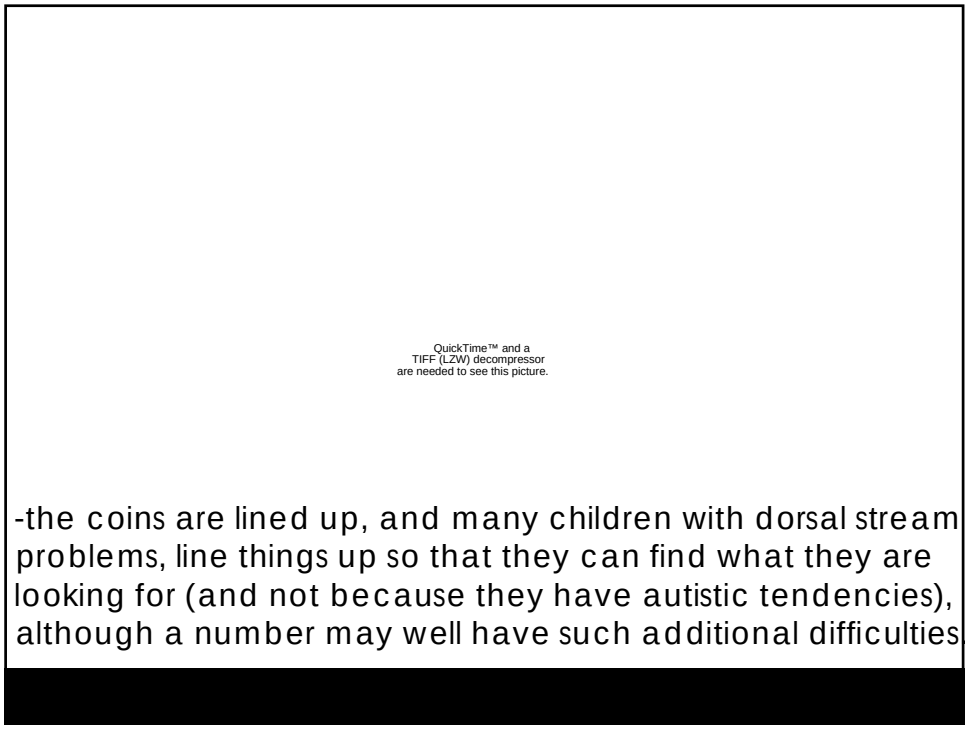
Difficulty seeing things that move, is common amongst children who have impaired dorsal stream function, and who often prefer films in which there is limited movement.

The principles of managing dorsal stream dysfunction are:

- Storing possessions for one, not three dimensional search
- Keeping background clutter and pattern to a minimum. (Unlike in this slide!)
- Limiting the amount to see at any one time



It is much easier to count coins or find a coin if -



-the coins are lined up, and many children with dorsal stream problems, line things up so that they can find what they are looking for (and not because they have autistic tendencies), although a number may well have such additional difficulties

Storing toys and clothes so that search is in one direction

QuickTime™ and a
TIFF (LZW) decompressor
are needed to see this picture.

And minimising background pattern helps in finding things

QuickTime™ and a
TIFF (LZW) decompressor
are needed to see this picture.

Children get close to the television to limit the amount they are seeing at any one time.

In the classroom this translates into:

- Having good storage and putting everything away
- Minimising the amount of decoration
- Allowing the child to get close to the front
- Ensuring that there is not too much on the page and that the separation of words and numbers is sufficient
- (Consider *san serif* text like this)

Additional strategies include:

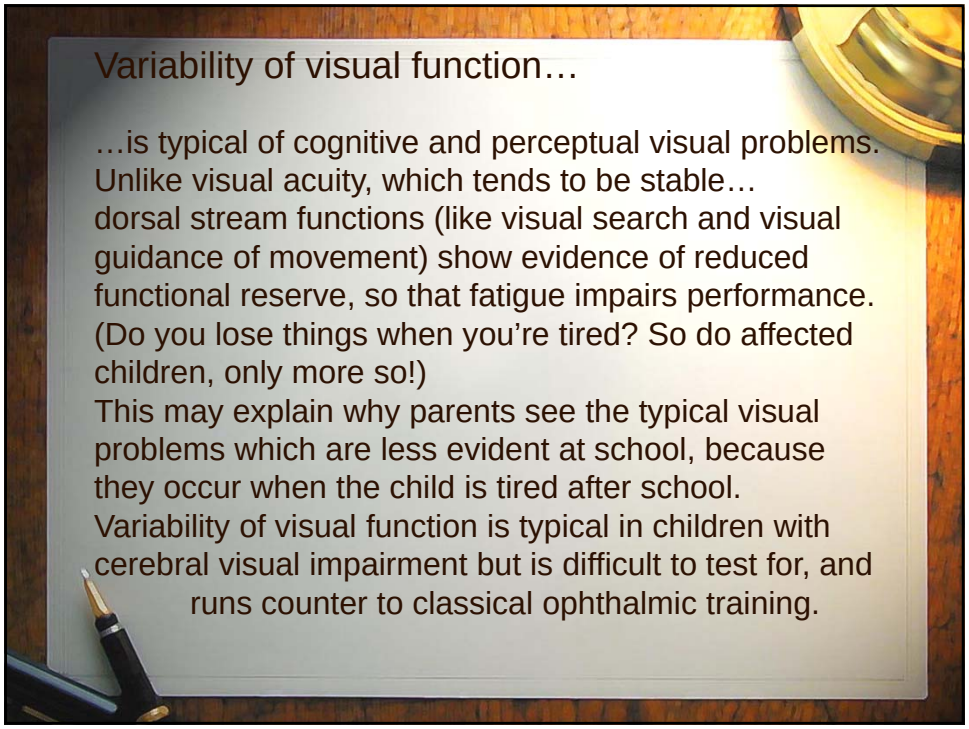
- Limiting all forms of distraction for the child
- Using a ‘quiet table’
- Choosing the ‘neighbours’ to be the most calm in the class
- Using large well spaced material for group presentation
- Matching the speed of presentation to the speed of processing

For writing and reading:

- Squared paper can be used to present arithmetic and other ordered data
- And providing squared paper to write on can assist in not losing the place, especially when dealing with columns of numbers

In the playground and outside:

- Recognising the social isolation due to not being able to see a friend in a moving group on the playground, and taking appropriate action, to ensure the child is integrated into the group and not standing at the side of the playground, unable to join in
- Choosing appropriate sports which the child can do well in if team activity is not possible



Visual Fatigue

- Occurs following prolonged visual processing
- Inattentive periods due to tiredness and reduced “functional reserve” can be helped by:
  - Minimising clutter
  - Keeping distraction to a minimum
  - Reducing detail and complexity
  - And giving well earned breaks, ideally in a calm undecorated, and non-crowded environment

Dorsal stream dysfunction affects attention

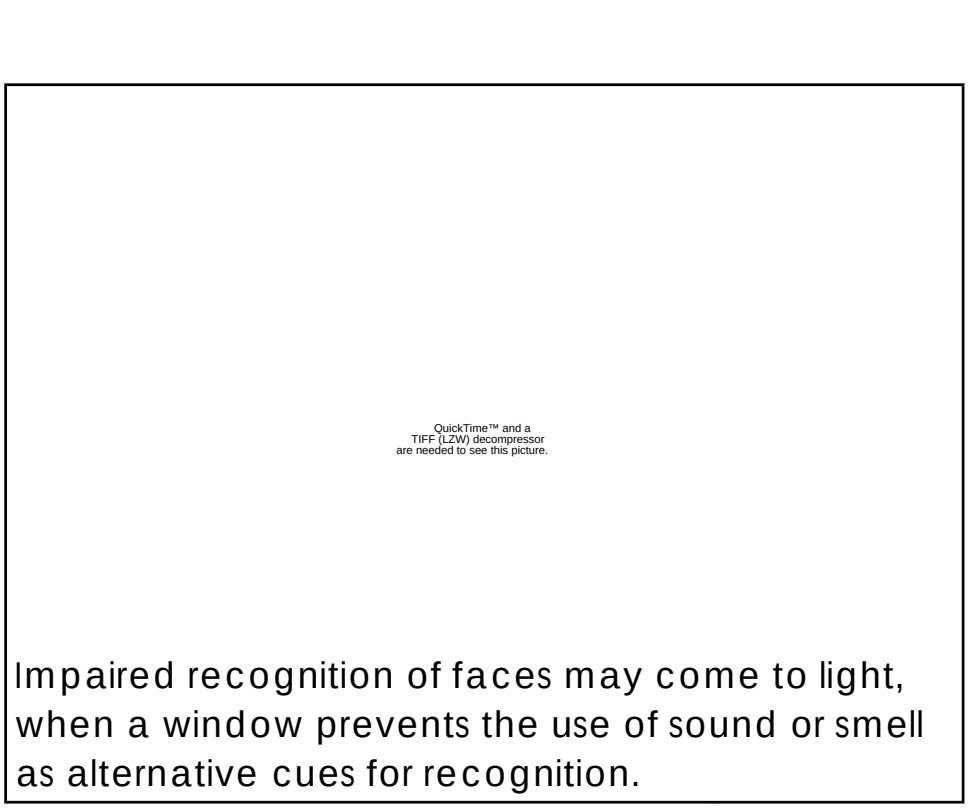
- It can be difficult to engage, to maintain and to break visual attention
- Shared attention during listening and talking can be very difficult to keep going. It is difficult to watch a face and listen to what is being said, so the child may look away. To say “Look at me when I’m talking to you”, can make the problem worse!
- There can be loss of visual attention when things are moving at the side, and it can be difficult to return to the work in hand.
- You too may have similar problems with this slide!

Behaviour

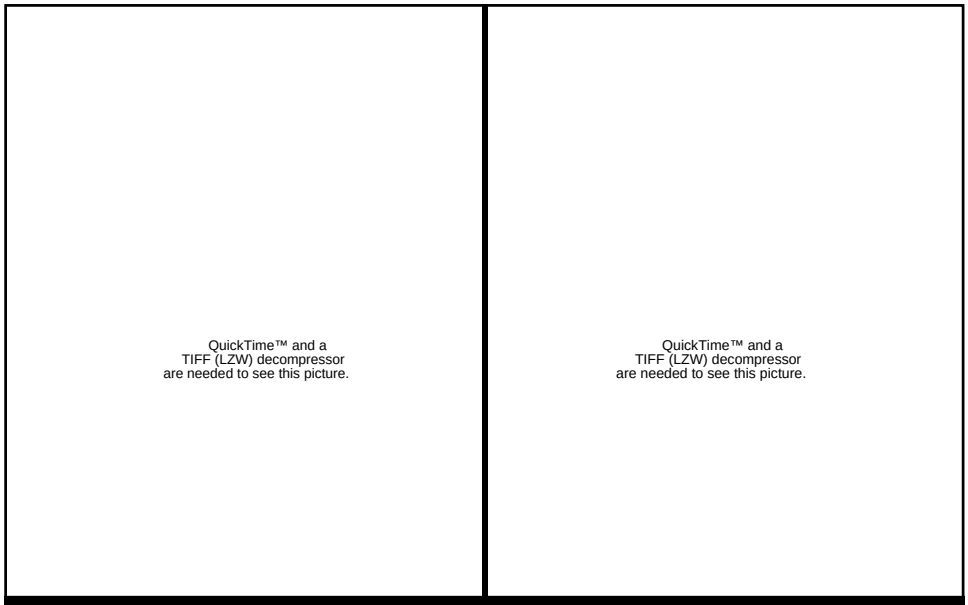
- Rooms with a lot of pattern and clutter, and being in crowds and supermarkets are associated with difficult behaviour or distress (Do you find the background under these words annoying?)
- Angry reactions can take place when other restless children cause distraction, particularly when a child is concentrating
- But plain undecorated places and quiet open countryside are associated with better behaviour (Isn't this lower text more relaxing to read?)

Ventral stream dysfunction - results in impaired recognition

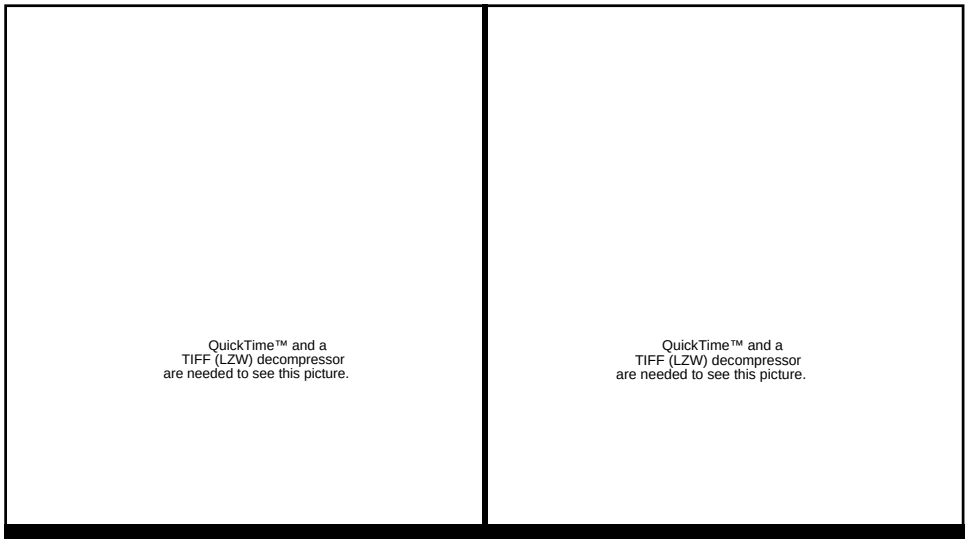
In the context of adequate visual acuities



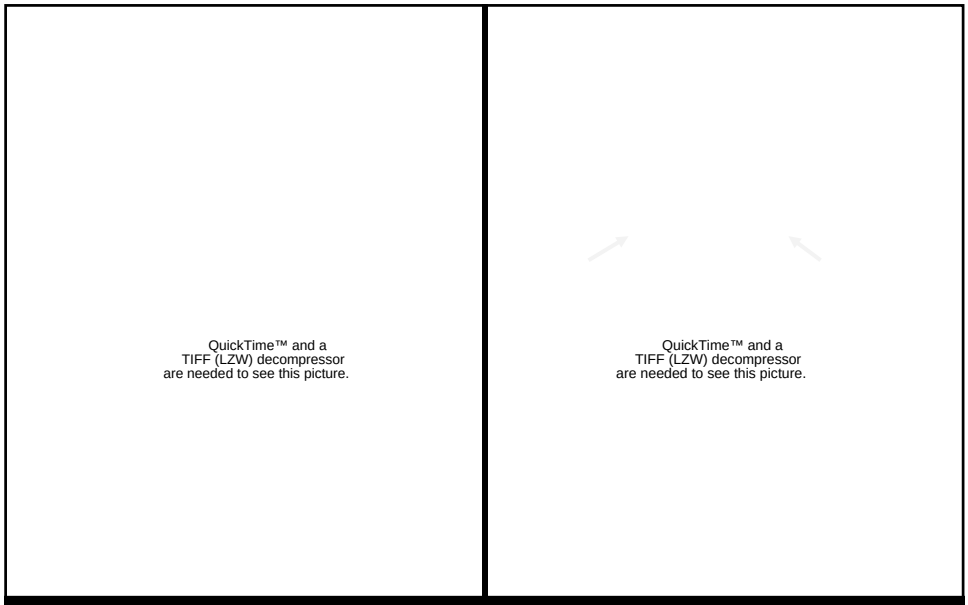
Impaired recognition of faces may come to light, when a window prevents the use of sound or smell as alternative cues for recognition.



In incomplete and milder versions of impaired recognition, side on views may not be possible to recognise, when square on, (or canonical) views are not a problem.

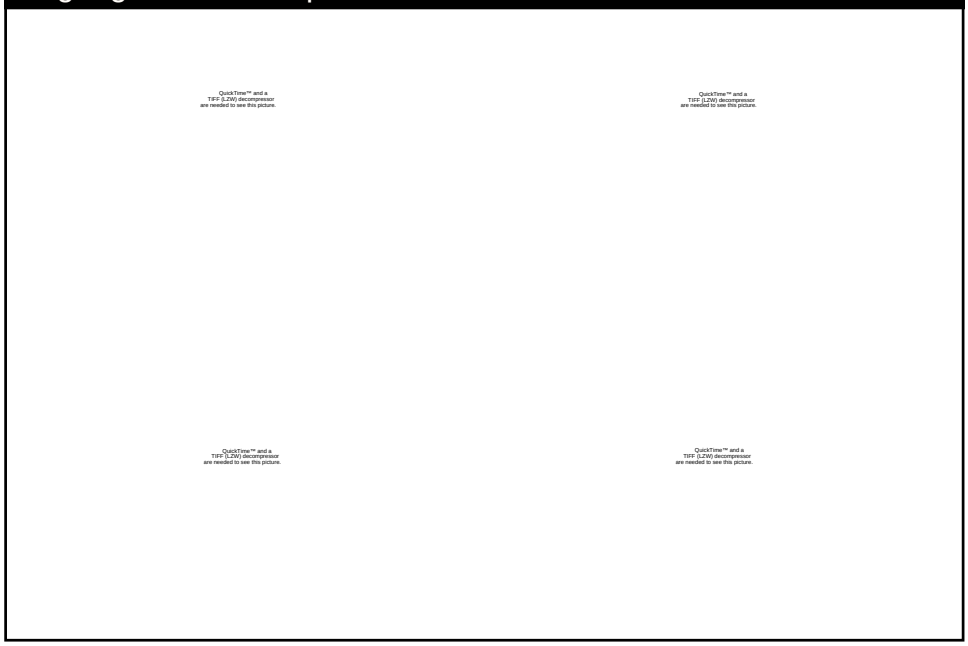


The act of not recognising someone who is not known is a complex 'computing' task, and children with impaired recognition commonly approach strangers thinking that they know them.



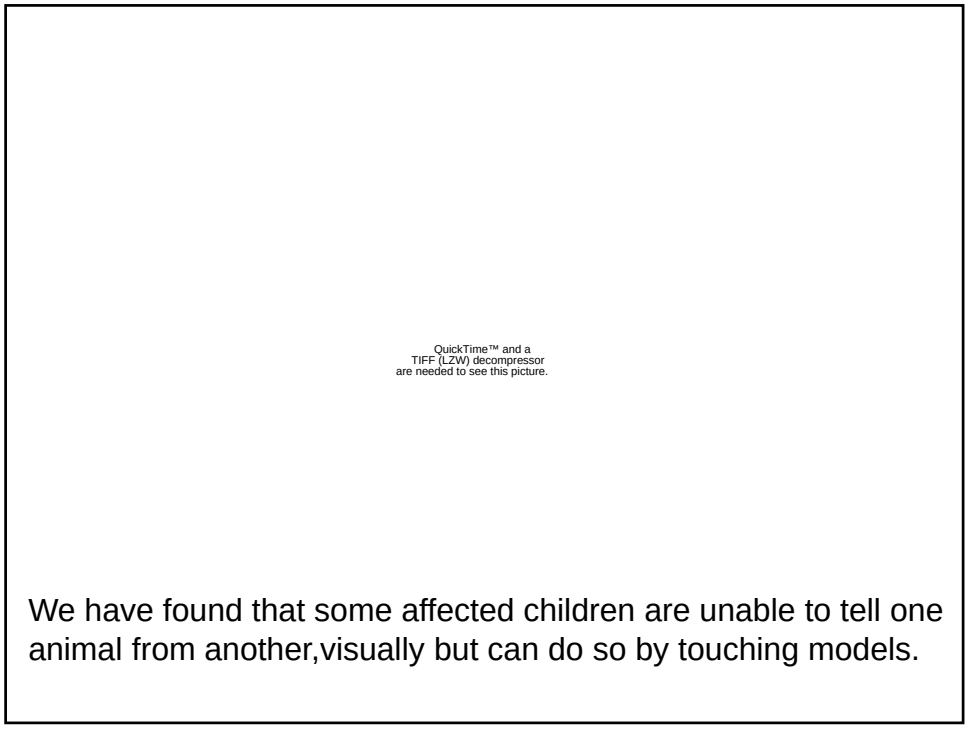
Remembering the colour of clothing and the shape of identifiers such as earings, and voice recognition are all strategies to circumvent the problem.

Many children who cannot recognise faces are unable to see the language in facial expression. As are those with low acuities.



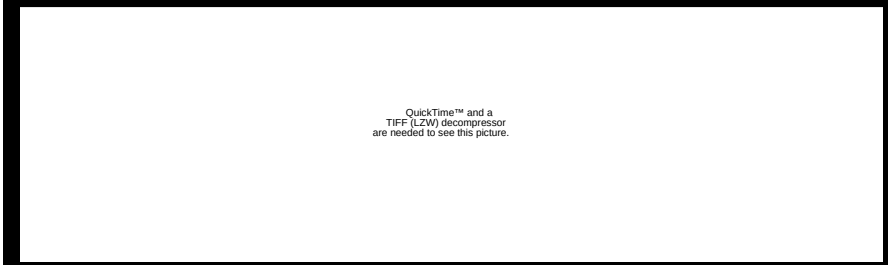
It is important this is recognised by asking children to identify facial expressions like this, or they may easily be misunderstood

Communication in which feelings and emotions are all made explicit is essential for these children, working on training about the meaning of the expressions.

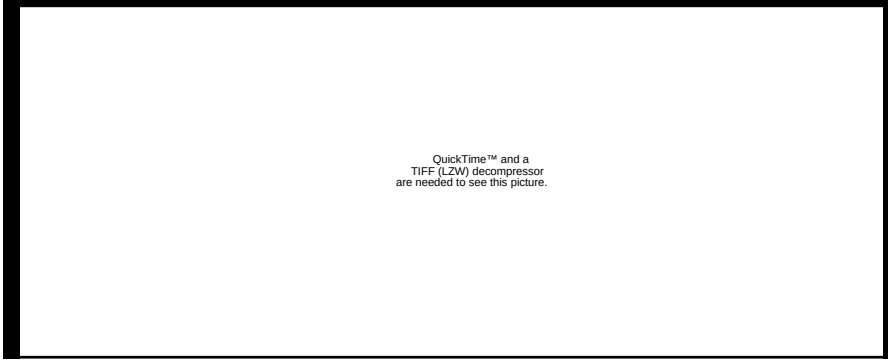


We have found that some affected children are unable to tell one animal from another,visually but can do so by touching models.

Object recognition may need to be enhanced by touch.



And the family car may only be recognised by colour.



QuickTime™ and a
TIFF (LZW) decompressor
are needed to see this picture.

And orientation at home in the garden and at school may be very difficult, and more profoundly disturbed than the orientation problems due to dorsal stream dysfunction, (where it is overload of the visual system which probably causes the problem.

Some families have found that color coding doors and routes can help considerably.

QuickTime™ and a
TIFF (LZW) decompressor
are needed to see this picture.

Temporal constraints in children with cerebral visual impairment

Communication with affected children must be perceptible & enhance what the child is giving attention to.

It therefore needs to be paced within the child's speed of processing.

The social problems due to CVI

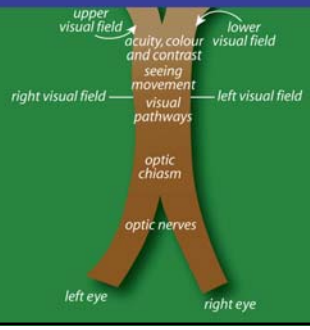
Problems	Approaches
• Labelled as having 'behavioural problem'	• School Support
• Self Conscious	• Identify problems and solutions • Encourage child to overcome them
• Feelings of isolation	• Well known peer group • Buddy system • Find activities child enjoys and can excel in

Conditions with cognitive and perceptual visual problems

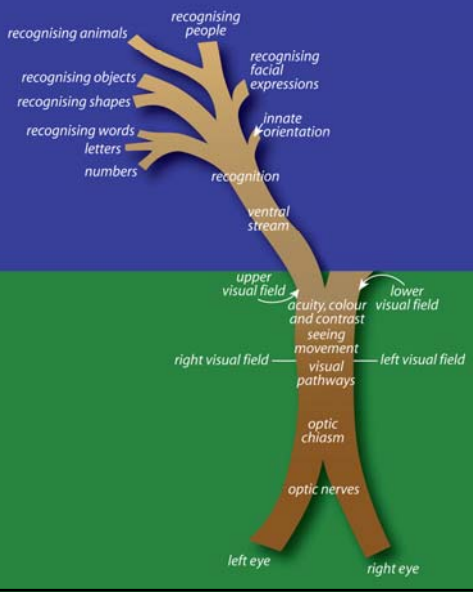
- Hydrocephalus (>50% of cases)*
- Occipital encephalocele
- After meningitis and encephalitis
- After head injury
- Vascular events
- Neonatal hypoglycemia
- Children with cerebral palsy
- Part of 'cortical' visual impairment

*Andersson S, Persson EK, Aring E, Lindquist B, Dutton GN, Hellstrom A. Vision in children with hydrocephalus. Dev Med Child Neurol. 2006; **48:** 836-41.

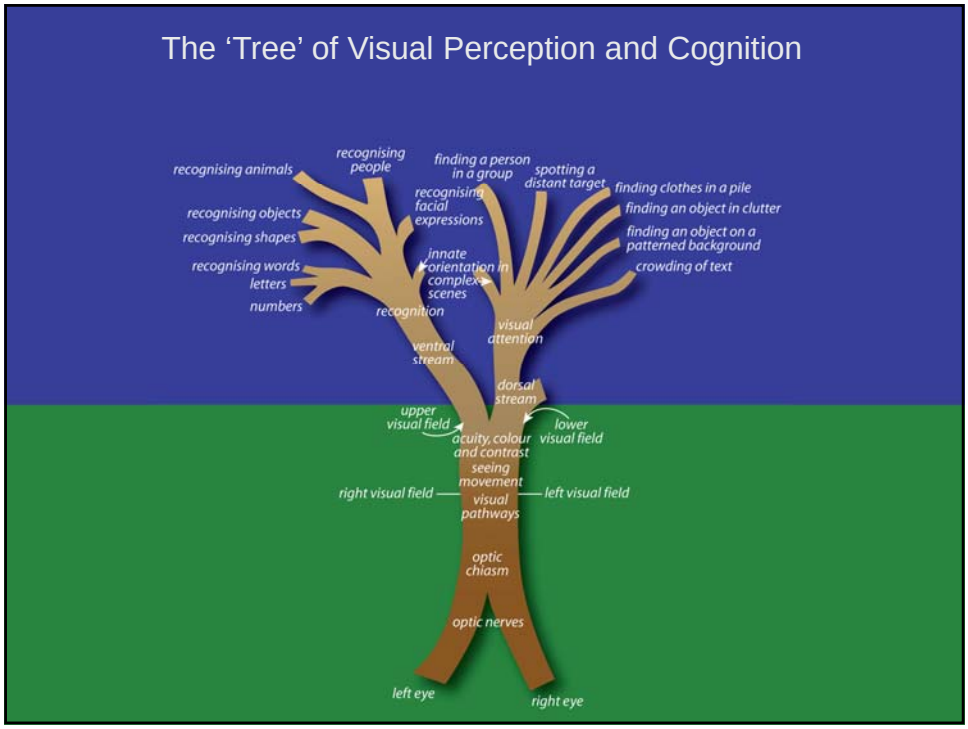
The ‘Tree’ of Visual Perception and Cognition



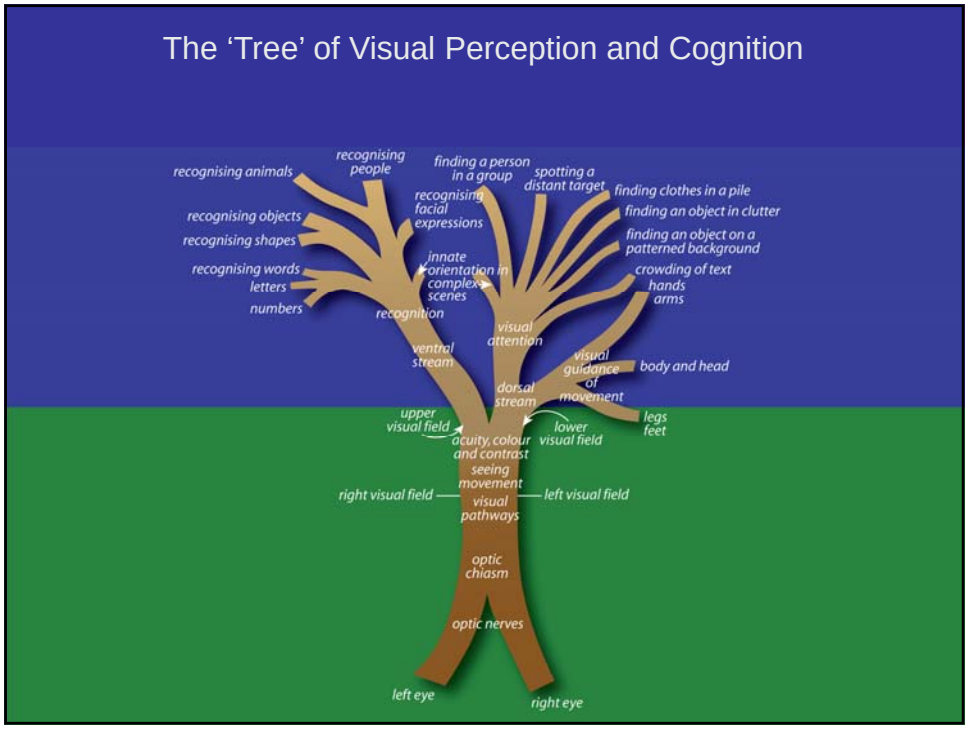
The ‘Tree’ of Visual Perception and Cognition



The ‘Tree’ of Visual Perception and Cognition



The ‘Tree’ of Visual Perception and Cognition



As an aside:

All of the problems described in this lecture affect adults as well.

Agnosia is not to know.

Anosagnosia is not to know that you don't know.

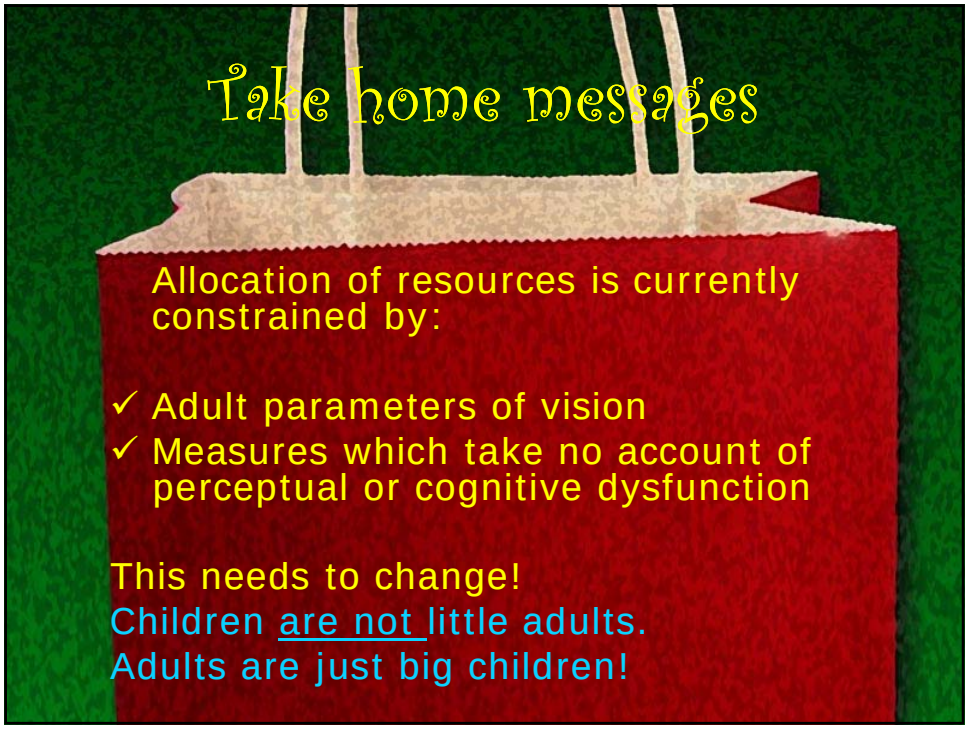
Adults with damage to the visual brain, often also have damage to their insight.

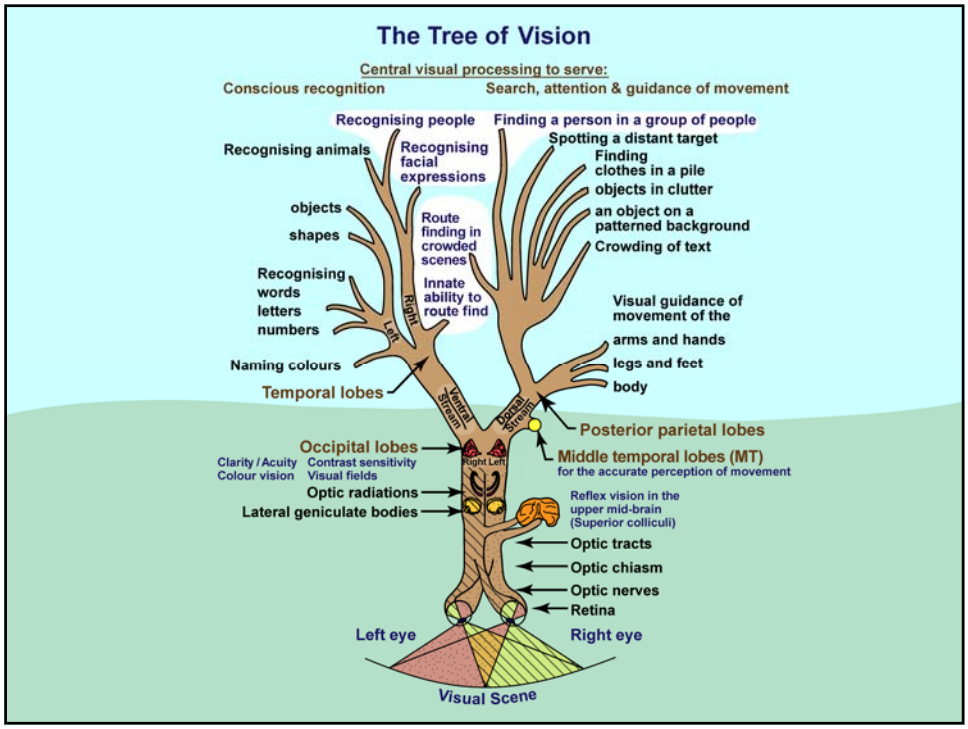
Thus like in children, their visual problems are frequently non-symptomatic!

In conclusion:

Disabilities due to CVI can be obscure , and are often missed. Yet the resultant problems may be disabling. Recognition and habilitation for this common disorder are essential, to prevent long term educational and social disadvantage of affected children

Take home messages
Vision is fundamental to optimal child development
✓ All aspects of impaired vision need to be identified, and acted upon, by staying within visual thresholds
✓ Visual acuity is often nearly normal in CVI, which is by far the commonest cause of visual impairment in children
✓ The change in attitude and approach which accompanies the understanding of visual behaviour can be life changing (for infants to teenagers).


Take home messages
Allocation of resources is currently constrained by:
✓ Adult parameters of vision
✓ Measures which take no account of perceptual or cognitive dysfunction
This needs to change!
Children are not little adults.
Adults are just big children!


The Tree of Vision
Central visual processing to serve:
Conscious recognition
Search, attention & guidance of movement
Recognising people
Finding a person in a group of people
Recognising animals
Recognising facial expressions
Spotting a distant target
Finding clothes in a pile
objects in clutter
an object on a patterned background
Crowding of text
objects
shapes
Route finding in crowded scenes
Recognising words letters numbers
Innate ability to route find
Right
Left
Visual guidance of movement of the
arms and hands
legs and feet
body
Naming colours
Temporal lobes
Ventral Stream
Dorsal Stream
Posterior parietal lobes
Occipital lobes
Clarity / Acuity
Colour vision
Contrast sensitivity
Visual fields
Right Left
Middle temporal lobes (MT)
for the accurate perception of movement
Optic radiations
Lateral geniculate bodies
Reflex vision in the upper mid-brain (Superior colliculi)
Optic tracts
Optic chiasm
Optic nerves
Retina
Left eye
Right eye
Visual Scene