

Understanding and managing children with profound cerebral visual impairment

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Cortical Blindness

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- A condition in which there is profound damage to the back of the brain which is responsible for vision, - the occipital cortex and the pathways leading to it
- This can be due to many causes. The commonest is lack of oxygen and blood supply causing a 'stroke', usually early in life

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- The visual fields are markedly constricted of typically hemianopic / non-attentive on one side
- But there is usually some detectable visual function

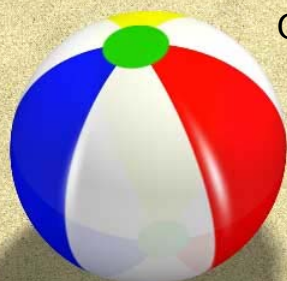
Cortical Blindness

At a basic level:

- The front of the brain thinks
- The middle moves and feels the body, and hears and interprets language
- And the back sees
- All three parts, or two parts or only the back (more rarely) may be affected in any combination

Profound Visual Impairment is the result, but this may be accompanied by apparent intermittent ability to detect movement, or “Blindsight”

(Which can be thought of as seeing without knowing)



We have two 'visual brains':

✓ A primitive reflex visual brain (which allows us to rapidly, and unconsciously avoid moving hazards. It at the top of the mid-brain at the back

(and is large in primitive animals like crocodiles)

✓ And our developed, cerebral visual brain, the occipital lobes at the back

Blindsight - there are two forms:

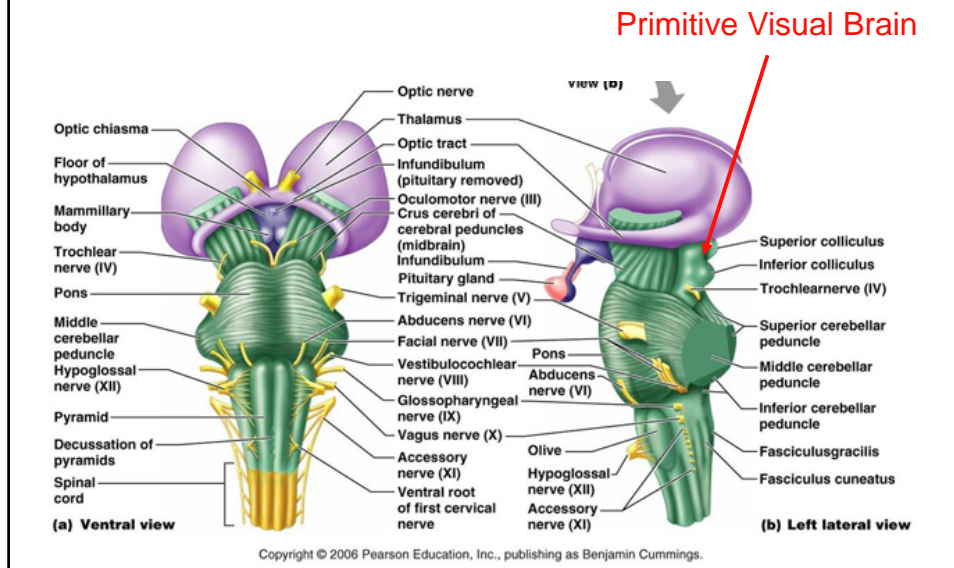
Low level blindsight or brain stem vision This is:

Reflex (as an automatic reaction)

Peripheral (only responsive through peripheral vision)

Fatiguable (happens once or twice before fatiguing)

This is a diagram of the brain stem and vision is served near the top at the back in the location shown by the red arrow.



Low level blindsight

- If you are driving along and a child on a scooter crosses the road in front of you, you use your automatic primitive blindsight system to prevent an accident

.....

QuickTime™ and a
H.263 decompressor
are needed to see this picture.

Blindsight

Low level: Brain stem vision

Reflex

Peripheral

Fatiguable

High level: Dorsal stream vision

This

form of vision:

- Allows us to move through the visual world
- Is unconscious but can be rendered conscious
- Is trainable in those who have lost vision due to visual brain damage (by giving confidence to use it!)

Hemianopic Blindsight

In a child with loss of vision on one side
blindsight may allow movement in the blind area to
be detected and reacted to. (In the next video you
can see a simulation of hemianopia you can see this
happening.)

QuickTime™ and a
H.263 decompressor
are needed to see this picture.

Patient MC

High level blindsight

Severe respiratory illness with very low blood pressure:

- 27 days unconscious
- Woke unable to see due to stroke to the occipital lobes of the brain
- Seen 2 years later

...when she looked at a window in the rain, she could see the rain drops, but could not see through the window!

QuickTime™ and a
H.263 decompressor
are needed to see this picture.

...she could see a flowing stream, but could not see the branches of the trees which were not moving.

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

... she could see her young daughter's pony tail move to and fro as she walked away, but could not see her daughter.

QuickTime™ and a
H.263 decompressor
are needed to see this picture.

...When she was investigated by functional Magnetic Resonance Imaging, it was found that her motion detection brain (the orange parts) was intact.

And this enabled her to walk freely through the world she could only see in a blurred way when moving!

She had 'high level' Blindsight!

So I recommended that she use....



... a rocking chair. This enabled her to learn to see better.

QuickTime™ and a
H.263 decompressor
are needed to see this picture.

Children with blindsight

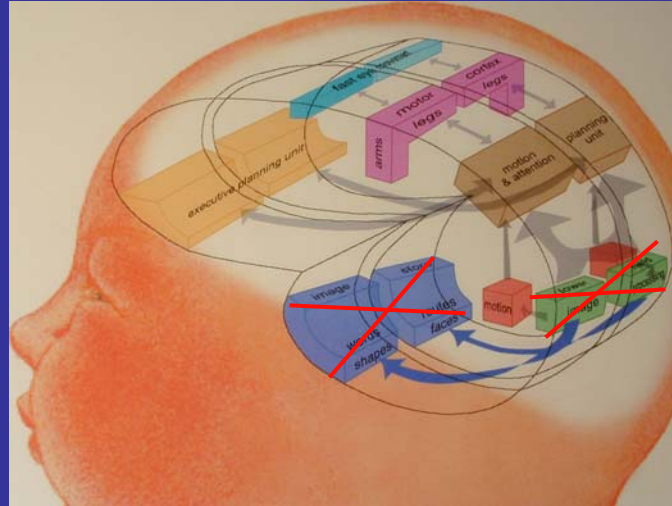
- Does this explain why some multi-disabled children who rock back and forth, do so, in order to gain some vision?

Returning to Patient MC

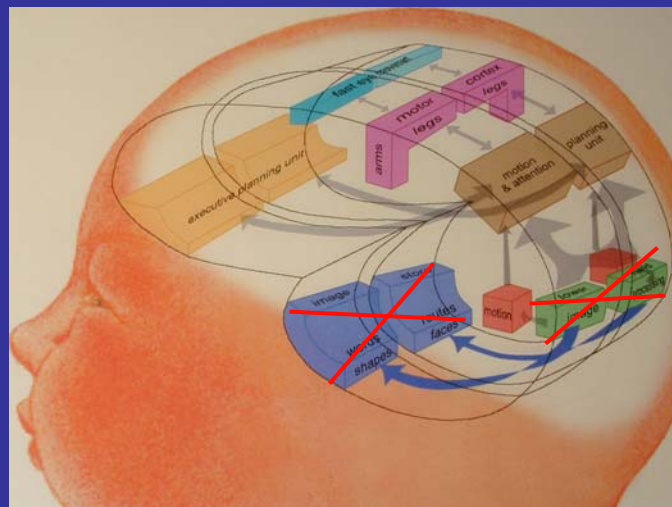
Despite perceiving movement only MC can:

- Walk freely around obstacles
- Reach for items accurately if they are moving, or she is
- Read letters at 20/80 (6/24) acuity by tracing her fingers over them

Although the centres (in the green and blue boxes in this diagram) are not working, the 'red and brown boxes', which serve movement detection and visual guidance of body movement still are.



We have seen a number of children with the same type of acquired damage to the brain who have been fully mobile, despite not being able to see anything, and being functionally blind!



For the common type of 'Low Level Blindsight' however, in quadraplegic children

Vision is best for movement to the side

Moving targets may be reached for at the side, but not straight ahead

Vision can be fatiguable and intermittent

It can vary from - day-to-day and from moment-to-moment

And the rare mobile child with 'blindsight' can walk slowly round obstacles.

This has been called travel vision.

Some of these children sometimes respond to smiles:

We know that adults with acquired cortical blindness can do this too.

This has been called: affective blindsight

And is due to the primitive visual brain, which has not been damaged, being able to respond to smiles!

So please believe it when parents of children with cortical blindness say that this happens. It does!

In profound cerebral visual impairment

- Some children see and perform better in dim lighting, probably because they have different brain settings for brightness - so use this lighting
- Find and use the best field of vision, and best location (with the child's normal comfortable head posture)
- Attention can only be given to one thing
- So ensure the child is comfortable without pain and-
- Make special events perceptible by being 'singular'

Speed of Processing in Severe damage to the Brain

- **For incoming information**
 - Speed of visual processing is slow
 - Speed of visual attention is slow
 - Speed of auditory processing - the words and the elements of the word
 - Speed of auditory attention is slow

Speed of Processing in Severe Damage to the Brain is Slow

- So that for performance as well as reception
 - The speed of perceiving
 - The speed of reacting
 - The speed of bringing about and the
 - Speed of completing are
 - All slow - which all adds up

And the **KISS** principle must apply

- Keep It Slow and Simple
- Consistently matched to and immediately following the experience of the child, as the child is having that experience
- Share the same vocabulary
- Repeat, and repeat and repeat

Recognise Blindsight behaviours

- Reaching for objects to the side but not straight ahead
- Looking away from people in order to see them
- The vision is fatiguable
- Vision may be intermittent on an hourly or even a day to day basis

Assessing the child Taking a history:

- The **open history** from parents - they know the child's abilities and what the child knows - make this the starting point
- The **structured history** to elicit:
 - Level of vision - what detail can be seen?
 - Field of vision - where does the child see?

Assessing the child Taking a history:

- **Structured history to elicit**
- **The timing of vision**
 - When is it best?
 - How long does this last?
- **Attention**
 - How much can be attended to?
 - What is known and what is favourite?
 - What drives and maintains attention?

Assessing the child Assessment of functional vision:

- **Make sure the child is comfortable**
- **Assess in a familiar environment**
- **Remove all distraction**
 - No clutter
 - Limited decoration
 - Limited noise
- **Provide a favourite toy, wait and watch**

Assessing the child

Assessment of functional vision:

- Is a face seen, and responded to?
 - If so, at what distance?
- Are large peripheral targets seen?
 - If so, in what quadrants?
- Is a central target seen and responded to?
 - If so, what was the response and how long did it take?

Principles to Consider

- Know that the child is 'normal'
- See, feel, hear and experience the world as the child does
- Ensure that everything is perceptible
- Give new experiences
- Enhance the experiences with consistent meaning, and optimal sequential timing
- Be a 'radio person' but not a 'television person'...

'Radio Language'

- Close your eyes and listen to the television. It does not make sense!
- But close your eyes and listen to the radio. It does make sense.
- When you use language use words that refer to the child's experiences as they happen....
-and use 'radio language' with no words which can only be understood by looking, because they refer to what is being seen!

For the sensory room

- Understanding that attention can only be given to one or two things
- Eliminate discomfort - create comfort
- Optimise posture -comfort helps attention
- Eliminate background sounds
- Use a singular visual stimulus - (not many)
- Provide a meaningful experience at the right speed
- Enhance the experiences one after another
- (Some children with profound CVI see better in darkened conditions)

The sensory room

- **Make each target -**
 - Visible
 - Meaningful
 - Interesting
 - A way of gaining, maintaining and prolonging attention
 - A route to learning
- **Learn the lessons & de-clutter sights and sounds elsewhere**

Thinking about the real world

- **At school and at home**
 - Inside
 - Out and about - city and country
 - Travel and its problems

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- At school and at home
 - Inside
 - Out and about - city and country
 - Travel and its problems
- **Supporting and teaching the parents & carers**
 - Thinking about space and time
 - Thinking about attention and its enhancement
 - Thinking about communication and language
 - Using blindsight for feeding

Travel Problems

Coping with distress in the car

- Distraction by listening to favourite music or stories
- Covering the eyes
- Darkened 'wraparound' lenses to decrease
 - Light levels
 - Moving / flickering visual stimuli

Is there a role for eye glasses?

For large refractive errors

- Children with cortical blindness may not accommodate (focus)
- Marked long sightedness may not therefore be automatically corrected
- Marked short sightedness common in prematurely born children

Uncorrected optical errors cause amblyopia which could add to the problem

Is there a role for eye glasses?

For large refractive errors

- If vision is much lower than would be caused by the refractive error - probably not
- If vision is a bit lower than would be caused by the refractive error - yes - because:
 - Delayed visual maturation can be very slow
 - Amblyopia may be contributing

Is there a role for eye glasses?

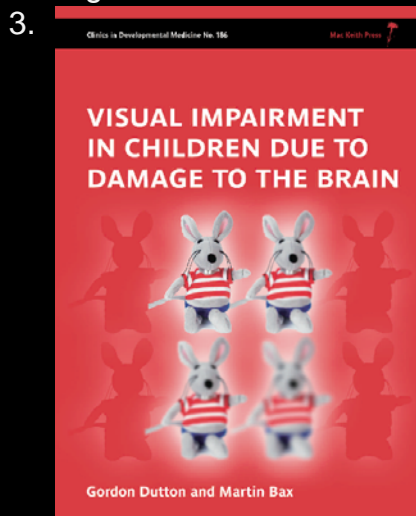
When hyoscine patches are used

- Hyoscine patches reduce salivation
- They can paralyse the pupils
- When they do, they also paralyse focussing (accommodation)
- Glasses may be needed because -
- Paralysed accommodation means:
 - No focussing for near
 - No focussing to overcome any long sightedness present

In summary

- Understand the limitations of brain processing
- Get into the mind of the child
- Capture the moment and enhance it with consistent repetition
- Give special time
- Recognise attentional limitations
 - In amount (in space)
 - And in time
- -and work within them
- Add to what you know the child knows
 - This is brain training!

1. Visual impairment Scotland Web-Site: VI Scotland
2. Sight-Sim web site.



4.

On-Line Course on
Cerebral Visual
Impairment
for Professionals
Working with
Affected Children

10 half hour lectures

**Emerald Education
Systems**

5. *Future publication*: Impairment of vision due to disorders of the Visual brain in childhood: A practical approach.
Eds: Amanda Lueck & Gordon Dutton (AFB Press).