Vision: what are we looking at?
A practical approach to assessing functional vision in children with cerebral palsy

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- **DVC**: Paediatrician, Psychologist, Occupational Therapist
- Assessment & management of severe visual impairment and its developmental impact
- Children with ‘isolated’ VI or in context of brain problem

- **CLT**: Speech & Language Therapy, Occupational Therapy, Paediatrician, Healthcare Scientist, Clinical Psychology
- Assessment & management advice for children with complex (physical) communication needs (CCN)
Vision: ‘just’ one of five human senses?

By Fabio Pantoja, via Deviant Art
Why think about vision in CP?

- Increased risk of visual disorders (inc severe VI)
- Ocular dysfunction AND brain dysfunction

- these difficulties must be systematically identified and addressed to optimise the child’s learning, interaction and development
Given the developmental challenges in CP: think functional vision!

- Where movement is severely restricted, vision has enhanced role for ‘information input’
- When controlled movements and vocalisations are limited, ‘information output’ achieved through functional use of gaze

THEREFORE

- Our observations of child’s patterns of gaze are informative as regards child’s capacity to initiate, respond (and TELL) within communication and learning situations
Vision in CP: what is said?

- VI – sees Specialist Teacher
- VI – under Ophthalmology
- VI – uses peripheral vision
- CVI – the world looks like swiss cheese
- Vision assessment: ‘fixes and follows’
- Vision: wears glasses, registered blind
- Visual impairment: wears glasses
Visual disorders in children with CP: reduced acuity

UK CP registers *Surman 2006*

- Any visual impairment: 34-40%
- Severe visual impairment: 9-11% (<6/60)

SCPE 2002

- Severe visual impairment: 11% (<6/18)
Visual disorders in children with CP: by CP subtype

Dyskinetic CP

• 11/48 reported VI
  \( (<6/18) \)

\( Himmelman 2007 \)

\( Himmelman 2009 SCPE \)

• DysCP: 242 (45%) VI, 19% severe VI \( (<6/60) \)
• BSCP : 45% VI, 38% severe VI
Visual disorders in children with CP: by CP subtype

Spastic CP  

Venkatswaran 2008

• 14 yr retr. case series; n=92, 39 prem
• 80% ‘visual impairment’ (n=83)
• Normal/mild-moderate/severe
• ‘Mild-moderate = squint, refractive error, amblyopia, nystagmus’
• ‘Severe = cortical blindness/optic atrophy’
Visual disorders in children with CP: by CP subtype or functional severity

Quebec registry study, 4y, n=301
Ascertainment btw 2y and 5y  

<table>
<thead>
<tr>
<th>Neurologic subtype</th>
<th>Cortical blindness (n = 23)</th>
<th>Severe visual impairment (n = 23)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Spastic quadriplegia (n = 85)</td>
<td>18 (21)</td>
<td>1 (4)</td>
</tr>
<tr>
<td>Spastic hemiplegia (n = 77)</td>
<td>2 (3)</td>
<td>- (-)</td>
</tr>
<tr>
<td>Spastic diplegia (n = 52)</td>
<td>1 (4)</td>
<td>1 (3)</td>
</tr>
<tr>
<td>Dyskinetic (n = 16)</td>
<td>1 (7)</td>
<td>5 (12)</td>
</tr>
<tr>
<td>Ataxic-hypotonic (n = 9)</td>
<td>1 (11)</td>
<td>13 (33)</td>
</tr>
</tbody>
</table>

Shevell 2009
Visual disorders in children with CP: case series: systematic assessment

Woo et al 2011  
- n=88 (84: spastic) GMFCS I-V  
  - 59% strabismus, inc vertical, 20% needed surgery  
  - 57 had acuity measured (Snellen)  
  - 24.6% corrected VA better eye ≤20/40  
  - 5.3% corrected VA better eye ≤20/200

Da Cuhna Matta 2008  
- n=123 GMFCS I-V  
  - fields, pursuit, strabismus, visuospatial neglect, acuity, nystagmus  
  - Acuity, n=100: 17 had uni or bilateral acuity loss (?details)  
  - 29/112 had eye movement problems  
  - 81 completed all tests, 38=47%, no visual disturbance, 28% 1 abnormality, 25% 2 abnormalities
Visual disorders in children with CP: systematic assessment

Ghasia 2008

• Cross sectional study, 50 ch with CP
• Representative of child CP popn
• GMFCS I/II: squint/amblyopia cf N child
• GMFCS III-V: ↑risk high myopia, absent fusion, dyskinetic strabismus, severe gaze dysfunction, optic neuropathy, CVI
Visual disorders in children with CP: by brain lesion: peri-ventricular leukomalacia (PVL)

- Children with/without severe motor dysfunction
- Reduced acuity, crowding, field defects, ‘visual perceptual-cognitive problems’ ‘delayed visual maturation’; nystagmus, ocular motility defects, strabismus

*Jacobson 1996, 2000*
Visual disorders in children with CP: by brain lesion: PVL with spastic quadriplegia  

- Acuity reduction
- Refractive errors
- Strabismus
- Oculomotor difficulties

- Acuity: ‘difficult to quantify’ assessment methods [include] observation....to detect ‘residual’ visual function

_Fazzi et al 2010_
Visual disorders in children with CP: by brain lesion: PVL and spastic diplegia Fazzi et al 2010

- Strabismus, refractive errors
- Optic atrophy or disc pallor
- Optic disc cupping
- Absent stereopsis
- ↓ acuity, mild/moderate
- Lower field defects
- Oculomotor disorders (?pure/ or attentional prob)

- ‘Visuocognitive disorders’
Cerebral Palsy: visual performance (& assessment responses) may be influenced by.....

- Strabismus
- Refractive errors
- Reduced acuity
- Oculomotor impairment
- Impaired visual attention
- Field defects
- Poor accommodation
- ‘Cognitive visual dysfunction’
- ‘Visuo-perceptual difficulty’

AND

- Neurodevelopmental status: motor/cognition
- Social impairment
- Alertness /motivation/ comfort
- Suitability of materials
Why assess eyes/functional vision?

• To identify problems that may be present
• To clarify whether such problems have a significant impact on function, or not
• To explore areas of parental (or professional) concern regarding eyes/vision and to clarify areas of misunderstanding
• To clarify whether abnormal functional visual responses are due to visual or non-visual difficulties: can be key to identifying significant cognitive impairment
Assessment: challenging but not impossible

‘it is often difficult to estimate visual f’n in children with profound neurological impairment, yet it is these children [for whom assessment is vital]’ McCulloch 2007

Evidence that all children with Special Educ Needs all require visual assessment: ‘with enough patience, [skill] & suitably adapted methods, assessment is feasible in most children with learning disabilities’ Das 2010

Ocular & visual status among children in special schools in Wales: the burden of unrecognised VI. 73/173 children never previously assessed. 6/166 acuity assessments: acuity too low to measure Woodhouse 2014
Visual detection: Acuity measures; visual fields

Visual cognition within wider developmental profile

Active use of gaze

Visual Recognition

Fixation/shifts; visual attention; search

Visual Function

MULTIPROFESSIONAL

the pyramid of assessment

OPHTHALMOLOGIST

Ocular structure & function

Ocular Health; Refraction; Accommodation; Strabismus and Ocular motility
But, assumptions may be made and things missed!

- Not all children have been seen at the Eye clinic
- Some have been seen – but what actually took place?
- Diagnosis previously given but ‘ignored’ as unwelcome

- Everyone thinks someone else identifies the significant problem
- ‘diagnostic overshadowing’: abnormalities ascribed to ‘CP’

- Lack of professional confidence in assessing a child with severe CP, additional impairments not taken into account
- Lack of our confidence in interpreting Ophthalmology letter

- And what about Cerebral Visual Impairment?
Eyes and vision: not the same thing

PARENT
‘they said his eyes were OK’

NON-EYE CLINIC PROFESSIONAL
‘the letters don’t say a lot about vision / what the consequences of the ocular/orthoptic findings are’
Seen in the Eye clinic?
What’s overlooked or not reported?

• Acuity measures – or why this is not given
• Was a ‘diagnosis’ given – and rejected?
• Qualitative comments on visual attention, or purpose of fixation
• Eye movement difficulty? Or attention?
• Impact of other difficulties: may be substantial
• Abnormal visual performance may not be due to ocular/visual difficulty
We need dialogue with our Ophthalmology colleagues

• Clearly describe any concerns you have about the child’s vision

• Write a structured referral letter, stating clearly what information is sought

• Consider carefully the information you receive after the consultation, and clarify further if necessary

• Are there any Ophthalmology colleagues who have or who can develop a particular interest in the needs of children with CP?
Visual assessment in the child with CP: approach to assessment

Normal, or abnormal vision?
Normal, or abnormal functional use of vision?
• What is abnormal?
• What factors may explain this?
  – Ocular factors
  – Developmental factors

What exactly do we assess?
<table>
<thead>
<tr>
<th>Age</th>
<th>Behavior</th>
<th>Development of visual skills</th>
</tr>
</thead>
<tbody>
<tr>
<td>0-4w</td>
<td>Blinks to flash/Turns to diffuse light: <strong>awareness</strong></td>
<td>Interest in faces&lt;br&gt;Eye contact, responsive smile</td>
</tr>
<tr>
<td>6w</td>
<td>Watches close adult f&amp;f 6.5cm dangling ball</td>
<td>Selective <strong>attention</strong> for near targets: ‘one thing at a time’&lt;br&gt;Early visual search; tracking <strong>Visual interest</strong></td>
</tr>
<tr>
<td>4m</td>
<td>Watches adult 1.5m</td>
<td></td>
</tr>
<tr>
<td>5m</td>
<td>Blinks to threat</td>
<td>Object <strong>recognition</strong>&lt;br&gt;Visually directed reach</td>
</tr>
<tr>
<td>6m</td>
<td>Watches adult 3m&lt;br&gt;Fixates 1.25cm object</td>
<td><strong>Attentional shifts</strong> between near-near and near-far targets&lt;br&gt;Gaze following&lt;br&gt;Supports joint attention</td>
</tr>
<tr>
<td>12m</td>
<td>Fixates 1mm object</td>
<td></td>
</tr>
</tbody>
</table>
What visual systems are developing?

‘Vision for doing’
- dorsal stream
- short memory
- allows automatic motor responses

‘Vision for knowing’
- ventral stream
- conscious
- memory store
Normal vision promotes/supports

LEARNING, INTERACTION AND COMMUNICATION

- Knowing what things are
- Knowing what things are called
- Knowing that things exist & how to locate in space
- Interaction with play partners
- People looking at things
Visual assessment in child with cerebral palsy: principles

START

Current questions

HISTORY

Previous findings

MIDDLE

OBSERVATIONS

CONCLUSION

As far as can be made on basis of current info
Ocular and vision history: including functional visual skills

- Previous Ophthalmological assessment – *full findings*
- Does the child wear glasses, purpose

- Vision history: elicit *descriptions* of child’s responses: history needs DETAIL
- Elicit any areas of parental concern: any factors in past history which could influence current beliefs?
- Beliefs about child’s ability and responses can arise out of hope and be influenced by information previously given (or not given)
Vision history: including current functional visual skills

• What does your child look at?
• What can your child recognise, and how do you know this? (objects, pictures, people)
• Does s/he show any difficulty with steady looking/need more time to look at things?
• Can s/he look steadily at an object so that you are sure what s/he is looking at?
• (Structured Functional Vision Questionnaire)
CP: Vision as ‘input’ and ‘output’

- Looking at target = exploration: what is it?
- Looking at target: that’s interesting, I like it!
- Looking at target: Get it for me, now!
- Looking at target: I want *this one* (not that one)!
- Looking at target: *that’s* the answer to your Q!

Gaze is *multipurpose*
The different functions of gaze need to be distinguished
Descriptions of gaze may need further exploration

‘sometimes she doesn’t look at things, I think it’s just her cerebral palsy’

‘he eye-points to what he likes’

She’s not looking at it as she’s got a visual field defect

‘he looks so quickly at his symbols, but if you miss it he won’t repeat himself’

He uses eye-pointing

his eye contact is terrible: we were told it’s his CP’
An extract from Functional Vision Questionnaire (available on request)

<table>
<thead>
<tr>
<th>Question</th>
<th>Never</th>
<th>Less than I would expect</th>
<th>As much as I would expect</th>
</tr>
</thead>
<tbody>
<tr>
<td>Does your child look directly at a toy/object when this is placed directly in front of him/her?</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Comments</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Is your child slow in looking at the object placed in front of him/her?</td>
<td>Yes</td>
<td>No</td>
<td></td>
</tr>
<tr>
<td>Comments</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Does your child need an additional prompt to look at the object or toy? (please tell us what, eg sound, movement)</td>
<td>Never</td>
<td>Sometimes</td>
<td>Always</td>
</tr>
<tr>
<td>Comments</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Does your child track the object if it moves in front of him/her (NB tracking can be achieved either with head or eye movements or a combination)</td>
<td>Never</td>
<td>Less than I would expect</td>
<td>As much as I would expect</td>
</tr>
<tr>
<td>Comments</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>If two or more objects are in front of your child, does s/he look at each toy in turn, inspecting each toy? This is sometimes called shifting gaze.</td>
<td>Never</td>
<td>Less than I would expect</td>
<td>As much as I would expect</td>
</tr>
<tr>
<td>Comments</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>If there are 2 or more objects in front of your child, is it easy to tell which object your child is looking at?</td>
<td>Never</td>
<td>Sometimes</td>
<td>Always</td>
</tr>
</tbody>
</table>
Observations

- Detection
- Fixation quality, fixation shifts: inspection
- Acuity
- Recognition
- Exploration/search
- Eye contact
- Communicative/interactive/cognitive use of vision: ‘functional gaze’
Making Observations: Say What You See

Wonko the Sane (Hitchhiker’s Guide)
• “a scientist must also be absolutely like a child. If he sees a thing, he must say that he sees it, whether it was what he thought he was going to see or not. See first, think later, then test. But always see first. Otherwise you will only see what you were expecting.”

Sherlock Holmes
• “It is a capital mistake to theorise before you have all the evidence”
Observing ‘looking’

• Remember the importance of considering child’s *quality* of fixation

• Need to separate out what child is assumed to see, from what s/he can show s/he can see

• Preferential looking methods of acuity assessment use a ‘forced choice’ modality – assessor is unaware of location of target, and makes judgement as to what child has fixated on – *use same principle in assessment*
‘Blinded spotting’ useful for assessment

“Looking towards the top-left...”

“Blind” to position of correct answer and also of range of options present

Tester
Aware of position of correct answer and therefore open to “bias”
**Functional Gaze Control in Young Children with Cerebral Palsy**

Young children with severe physical disabilities who have little or no functional speech can rely almost entirely on their looking skills to engage with the world around them, interact with others and learn.

Observation of these children’s spontaneous and responsive gaze behaviours can be an effective way of assessing understanding of language, learning and participation in activities.

However, for observation of gaze behaviour to be of clinical value as an indicator of performance, clinicians first need to determine the child’s core repertoire of functional gaze control skills.
The assessment procedures are freely available to clinicians and families. We simply ask that you notify the **Functional Gaze Control team** to register your interest in these activities.
At the end of the rainbow......

Detect, resolve detail
Recognise what is seen
Maintain /direct /shift fixation: quality of visual attention
Distinguish one target from another: ‘choose’
Can target of fixation be ‘read’?

Overall: what vision is available for ‘exploration’, learning and communication

Is visual performance consistent with level of acuity/neuro-developmental profile?
Is child using controlled gaze to interact /respond?
Could any visual ‘difficulty’ be explained by any other factors?
Avoiding the pitfalls.....

- drawing premature conclusions
- relying on previous opinions
- ignoring the developmental profile
- distinguishing random from responsive movts
Are we missing ‘complex’ visual problems in children with CP?

Is it cerebral visual impairment?
Cerebral visual impairment: definition(s)

- ‘Loss of vision 2° to injuries to both geniculo-striate pathways’
  
  *Jan et al 1987*

- ‘Suspect cortical visual impairment in any child with ‘decreased vision’ and an otherwise normal ocular examination including normal pupillary light responses’
  
  *Lambert, Paed Ophth 1997*
CVI: newer definitions?

• ‘Severe impairment of vision due to cerebral cortical damage’
  
  *Dutton et al 1996*

• Decreased acuity, abnormal neurological imaging and abnormal electrophysiological testing consistent with bilateral damage to posterior pathways including the occipital lobes’
  
  *Good et al, Dev Med Ch Neurol 2001*

• Severe impairment of vision due to cerebral cortical damage.. A search for evidence of visual-perceptual impairment will usually be rewarded.......will lead to the implementation of a wider range of rehabilitative strategies
  
  *Dutton 1996, Dutton & Bax 2010*
CVI: so what do we mean?

• “When looking at visual dysfunction caused by brain damage or a brain development disorder, the medical term [CVI] is often used....Commonly, CVI includes all visual dysfunctions caused by damage to, or malfunctioning of, the retrochiasmatic visual pathways in the absence of damage to the anterior visual pathways or any major ocular disease

• One of the main reasons that consensus has not been reached [on who to describe as having CVI] is the heterogeneity of causes and symptoms which can be included”

Boot F, 2010
CVI: an *umbrella* term

- Group of disorders, with normal/reduced VA
- AND potentially accompanied by various additional consequences, not all present in each child
- Consequences may not manifest/be detectable until certain age
- In any individual, consequences may be of variable degree and impact may vary according to other co-morbidities, age, developmental skill level
- *Benefits from multidisciplinary approach to assessment and management*
Identifying CVI in CP?

• Assess the child in front of you

• Multidisciplinary step wise approach as previously described
Structured question inventories?

• Glasgow: Vision assessment clinic: Dutton et al
• Estimation of visual function difficult owing to communication problems
  – Infants/young children judged to have poor vision
  – Children with multiple disabilities
• Apparently paradoxical visual behaviour for which explanation & management sought
• ‘Visual skills inventory’
  – 6/36-6/60 or better

Dutton 1996 McCulloch 2007
Symptom patterns: ‘Cognitive visual dysfunction’

Questions to elicit impairment in:
• Recognition
• Orientation
• Depth perception (location in space, steps, floor surfaces)
• Perception of movement
• Simultaneous perception (sees part not whole)

CAUTION: Hearing impairment, Intellectual impairment, Autistic spectrum disorders, Difficulties recording acuity, Motor impairment

Structured history taking for children with evidence of CVI & measurable acuities facilitates identification, further investigation, diagnosis AND contributes to Mx plans

_Dutton 2010_

“Question inventories can be employed in the form of a questionnaire for the purposes of validation; but thereafter, their use is enhanced by expert history taking, in which clarification is sought to fully characterize the visual difficulties and their impact”

_Dutton 2011_
Visual skills inventory: key Q (???)

Does your child have difficulty in
• walking downstairs?
• seeing things that are moving quickly?
• seeing something that is pointed out in distance?
• locating an item of clothing in a pile?
• copying words or drawings?

_Could other factors influence responses? Are the questions relevant to children with CP?_

• _Careful clinical history to establish strengths and weaknesses tailored to child’s age, skill level and motor ability is the best start point._
CVI: think carefully

Van Genderen et al. 2012

• Short CVI ‘screening’ questionnaire
• 30 ch w CVI, 23 without CVI
• Visual and neuropsychological assessment

• ‘CVI questionnaires should not be used for screening purposes as they yield too many false positives
Assessment of cerebral visual impairment?

SYSTEMATIC ASSESSMENT APPROACH

• Systematic history & observations: gain information
• Assess the individual child in front of you: start from where this child is (not from where someone else thinks they are)

• it is only by thinking rationally, closely observing the child in front of you, and asking careful questions – running experiments – that you will be able to decide which explanation or hypothesis best fits your observations
Also...

- Cognitive assessment: verbal + non-verbal skills (WISC-IV)
- Picture detail and whole – scene with details
- Visual perceptual testing (e.g., DTVP-2)
- Face recognition
- Object recognition, picture recognition
- Motor coordination/planning Movement ABC or Bruininks
Higher visual processing skills

- Visual spatial/visual constructional /visual memory/visuo-motor integration—eg NEPSY II (arrows, block construction), Rey Osterreith Complex Figure, posting activity
- Visual attention: selective attention — TEA-Ch (Map Mission, Sky Search)
- Visual executive function — Delis Kaplan Executive Function battery (Trail Making, Colour Word Interference)
Case 2: A, aged 10y

- Bilateral dystonic CP, GMFCS V, MACS V
- 32wks gestation, ‘blood on brain’, abnormal CT
- Severe learning difficulty – understands key words

- Further advice on functional vision please
- Very variable responses observed; uncertain whether he can see what we show him
- Using ‘auditory scanning’; strengthening yes/no responses
- No particular family concerns about vision
‘Vision’ history

- No particular concerns about vision when younger
- Seen in an Eye clinic ‘somewhere’

- At nursery age wore glasses
- By 6y discontinued- ‘eyes corrected themselves’
- No concern raised by school staff

On asking directly what happened in the Eye clinic
- “The person who saw A showed him picture cards but he was not very interested in looking at them; the cards were grey and not colourful”
Current skills

• No interest in books - vision or concentration
• Notices animals moving
• Enjoys TV: head is at steadiest in front of TV
• Comic clips TV show: laughs at right moment; grumpy if sound off
• If on floor, cranes head round toward TV

• Communication: responds to choices offered
• ‘tunes in’ to some spoken words
Paediatrician refers to Ophthalmology: ‘concerns about functional vision’

“A report was not generated but clinic notes were reviewed .... the [ophth] team were ‘unable to answer the question ....about functional vision’. However there was ‘nil structural concern about the eyes.’ It was also noted that staff were ‘unable to complete Kays or Cardiff assessment. Ocular movements were variable, ? vertical gaze palsy, gaze upwards, doesn’t really fix/follow’.”
Referred again to Ophthalmology

- ‘visually inattentive, does not show central fixation. Appears to attempt to grasp visual targets’
- Fundus: very pale atrophic discs, retinas attached
- Variable convergent squint
- Refraction normal for age; rest of exam normal

- ‘significant amount of cortical visual impairment and optic atrophy’
- Now registered partially sighted
All clear now??

- Mother: what does this mean?

- School: so can we use pictures with him or not?
Further investigations?

• Visual evoked potentials: very poor responses seen: confirms clinical impression of low level of acuity and limited functional visual skill either for ‘input’ or ‘output’
• Meeting with school staff – be aware of tendency to assume eye direction indicates ‘seeing’; maximise opportunity for touch
Ocular and functional visual assessment in CP

• Important, but possibly not been done
• You can play a role in such assessment
• Functional vision includes detection, fixation, recognition, search etc
• Once assessment is done, SAY SOMETHING SENSIBLE
• Justify your conclusions and state what is not yet known
• Encourage Ophthalmologists to develop an interest in children with CP

THANK YOU
• Surman 2006 J Public Health 28(2) 148-156
• Himmelmann 2007 DevMed 49 246-251
• Himmelmann 2009 DevMed 94 921-926
• Venkateswaran 2008 DevMed 50: 216–222
• Shevell 2009 Neurology 2009;72;2090-2096
• Woo 2011 Optom Vis Sci 88 (12) 1520–1523
• Da Cuhna Matta DevNeurorehab 11(2) 159–165
• Ghasia 2008 InvesOphVis Sci 49(2) 572-580
• Jacobson 1996, DevMed 38 724-735
• Fazzi 2010 CDM 186 194-204
• Das 2010; Arch Dis Child 95:888–892
• Woodhouse 2014 Arch Dis Child 99:500–504
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• Dutton 2003 Eye 17 289–304
• Dutton 2011 DevMed 2011 53 389–393
• Van Genderen 2012 Strabismus 20(2) 78–83