The Changing Face of Cerebral Visual Impairment

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Children with CVI in mainstream schooling

- 82% have difficulties learning
- 91% have difficulties with friendship
- 100% need emotional support
- 100% have a high level of anxiety

McDowell; 2023

Overview

- What is CVI
- Prevalence and risk factors
- Population overview
- Assessment
- Management

Cerebral Vision Impairment (CVI)

- Vision impairment due to brain damage

- Reduction in **lower order visual functions**:
  - Visual acuity
  - Contrast sensitivity
  - Visual fields

- Reduction in **higher order visual functions**:
  - Visual perception (interpreting visual information)
  - Visual cognition (combining vision with prior knowledge)
  - Visual guidance of movement
  - Capacity to choose to give visual attention

- Can occur in conjunction with ocular vision impairment, but often the eyes are normal

CVI terminology

CVI - Cortical Visual Impairment

- The older term
- Still widely used in America

CVI - Cerebral Visual Impairment

- The newer term
- More commonly used in Europe
- Changed to reflect the fact that damage in any area of the brain can result in vision impairment, not just the visual cortex

Prevalence

- The most common cause of visual impairment among children in developed countries[1]
- One in 30 children in mainstream schooling[2]
- Prevalence has increased with improvements and developments in neonatal care
- For every child who is diagnosed with CVI, four more likely cases go unaddressed[3]

1. Gorrie, 2019; 2. Williams, 2021; 3. Perkins school for the Blind

People most at risk of CVI

- Cerebral palsy (60-70%)[1]
- Other neurodevelopmental disorders including autism
- Down syndrome(40%)[2]
- Other genetic or chromosomal disorders
- Ex-premmie babies
  - especially with very low birth weight and/or birth complications
- Developmental delay

[1] Philip et. al 2020 [2] Wilton et. al 2021

OTHER CAUSES OF CVI

Damage to the brain

- Closed head injuries
- Hydrocephalus, esp. blocked shunts
- Focal damage to specific brain locations due to haemorrhage, tumours, cortical dysplasia
- Metabolic disorders

Childhood damage to the brain

- Hypoxic ischaemic encephalopathy
- Neonatal hypoglycaemia (low blood sugar)
- Injury to periventricular white matter
- Meningitis and encephalitis
- Infantile spasms and epilepsy

Brain malformation

- Anatomical disorders of the visual brain
- Brain malformations
- Chromosomal and other genetic conditions

Dutton; 2015

Population overview: 3 functional groups – group 1

Children with profound disabilities and CVI

Disabilities impact on:

- Quality of vision
- Ability to move independently
- Cognitive abilities

This group may be:

- Unaware of things going on around them – appear non-responsive
- Unable to communicate thoughts and feelings to others
  - Often non-speaking / minimally verbal
  - May use a communication system e.g. PODD / PECS

Population overview: 3 functional groups – group 2

Children with CVI and further disabilities, but normal cognitive function

- e.g. A degree of CP, requiring assistance with walking, finds speaking challenging, but intellectually normal/high functioning

Normal vision when assessed with standardised vision assessments, BUT:

- Difficulties with visual guidance of movement
- Simultagnosia – seeing the whole scene at once
- Visual complexity

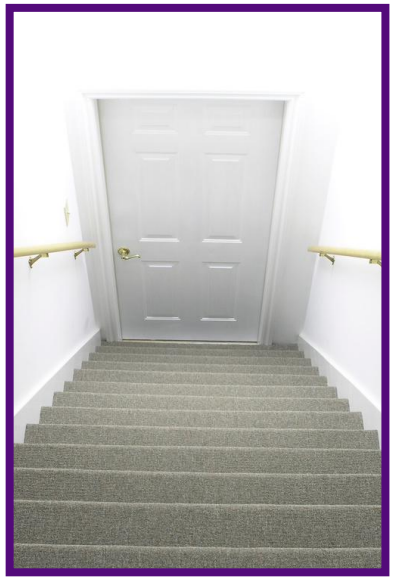
Dutton 2015

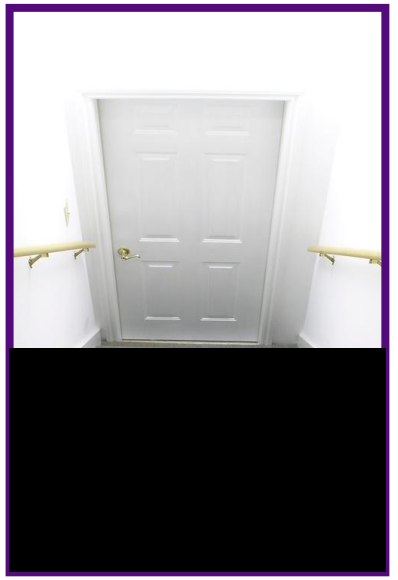
Population overview: 3 functional groups – group 3

Children with CVI and no other challenges

- Acuity range: very low to normal vision on standardised measures
- Visual field defects common, particularly lower field
- Perceptual difficulties possible:
  - Difficulties recognising objects, esp. viewed from atypical angles
  - Difficulties recognising people: social embarrassment
    - Possibly mistaken for autism or ADHD

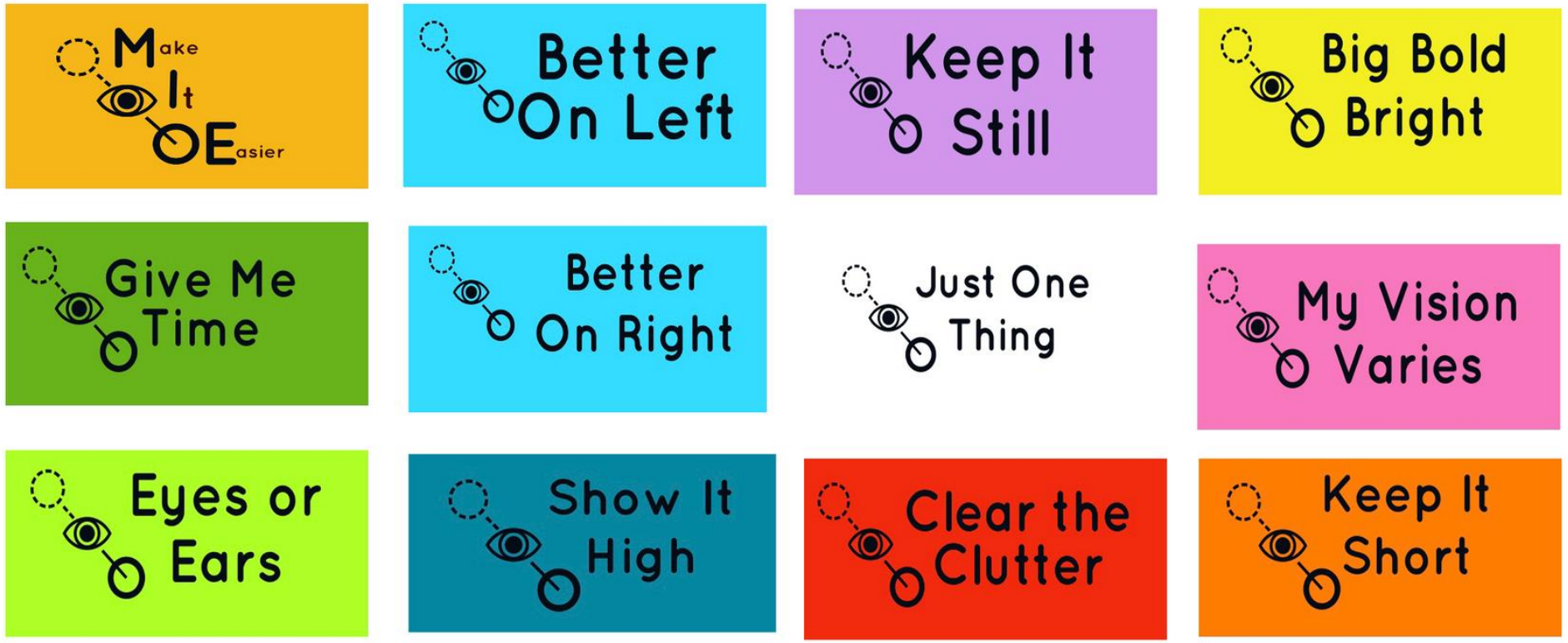
Difficulties often go unrecognised and the person develops coping strategies, often resulting in social isolation





Dutton 2015

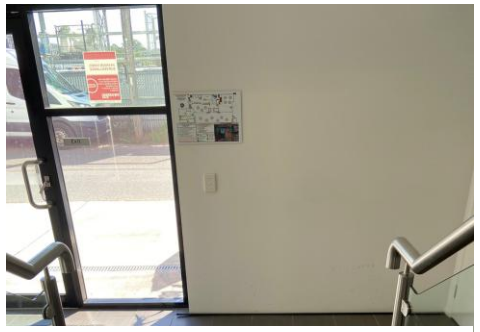
CVI - Functional Characteristics



Pilling; 2023

FUNCTIONAL CHARACTERISTICS - LOWER FIELD DEFECT

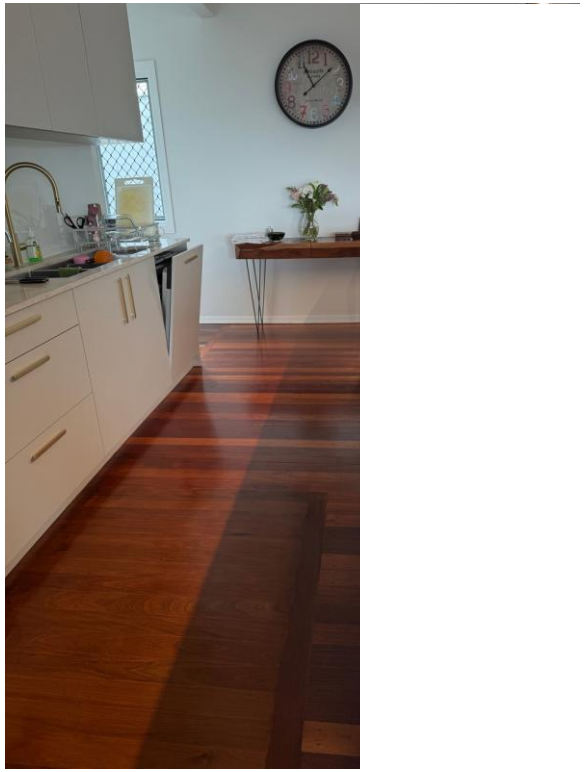
- Has difficulty walking downstairs
- Trips over/walks through obstacles on floor
- Get's 'stuck' at top of slide
- Gets angry and/or bumps into low furniture when moved



Show It High

FUNCTIONAL CHARACTERISTICS - HEMIANOPIA

- Leaves food on the right / left side of plate
- Bumps into doors / frames
- Misses pictures / words on one side of page



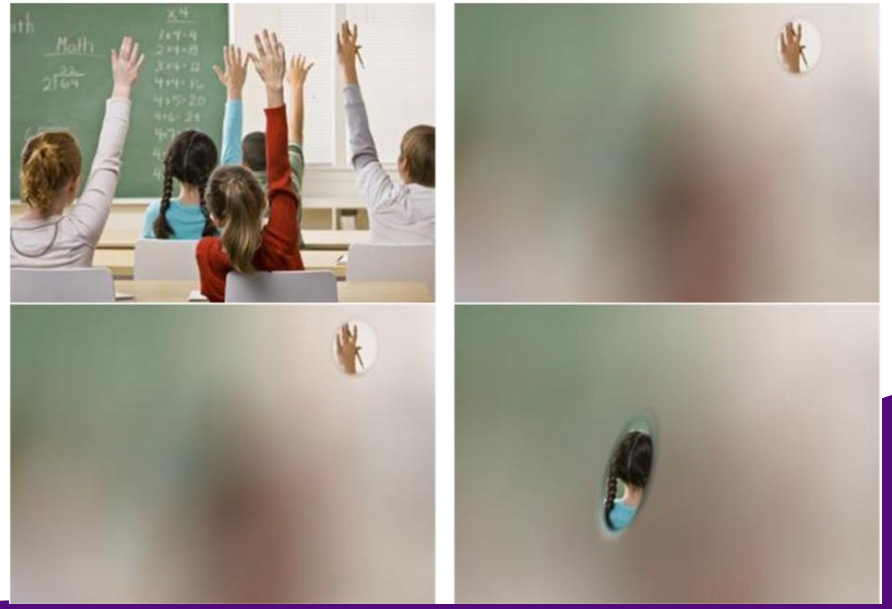
FUNCTIONAL CHARACTERISTICS - DUAL SENSORY PROCESSING ISSUES

- Bumps into things when walking and having a conversation
- Difficult behaviour in noisy / busy environments
- Appears unable to listen when visual engaged or not to see when listening

Eyes or Ears

FUNCTIONAL CHARACTERISTICS - SIMULTAGNOSIA

- Gets lost in places where there is a lot to see
- Cannot find friends in the playground
- Difficulties copying from the board



Used with kind permission from CVI Scotland

FUNCTIONAL CHARACTERISTICS - COMPLEXITY

- Seeing something pointed out in the distance
- Finding a toy in a toy box
- Finding people in a crowd

Clear the Clutter

FUNCTIONAL CHARACTERISTICS - MOTION PERCEPTION

- Difficulties seeing scenery from a moving vehicle
- Does not like watching fast moving TV
- Grasps incorrectly when reaching for objects / misses / knocks over
- Catching a ball

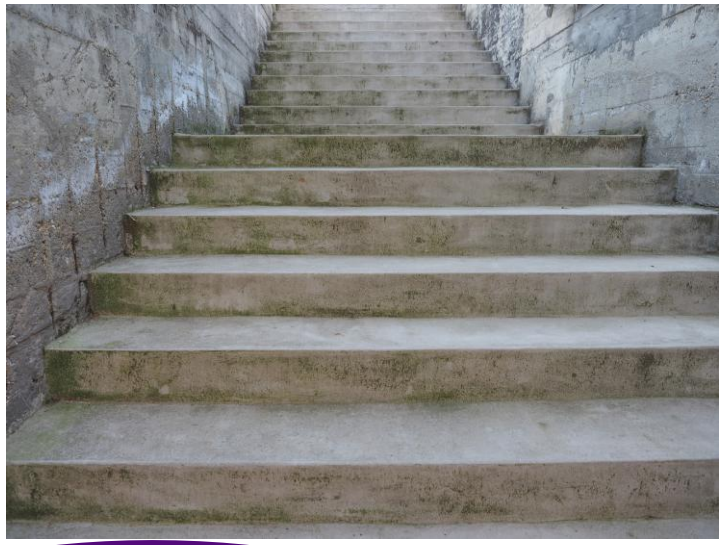
Keep It Still

FUNCTIONAL CHARACTERISTICS - LATENCY / FATIGUE / VARIABILITY

- Slow to visually interact with things
- Requires longer to find details
- Short span of visual attention
- Seems better able to visually attend sometimes than others

FUNCTIONAL CHARACTERISTICS - REDUCED ACUITY AND CONTRAST SENSITIVITY

- Difficulties seeing things when the light or contrast is poor
- Cannot see detail or clarity of vision



Big Bold Bright

FUNCTIONAL CHARACTERISTICS - OTHER

- Recognising faces and facial expressions
- Naming colours and shapes
- Recognising familiar objects e.g. family car

Diagnosis is challenging

- No standardised diagnostic tests/procedures
- Routine vision assessments: results within normal range
- No consensus on what constitutes a diagnosis
- Disciplines involved in diagnosis varies from country to country
- No established pathway to diagnosis in many countries, including Australia
- Huge range of function: assessment requires an equally broad range of assessments/approaches

Pathway to standardised assessment of CVI

2021 Review article explored assessments used to investigate and diagnose CVI in children

Conclusions:

- Lack of common practice in approaches to assess and diagnose
- Diagnosis of exclusion remained most common means to diagnose
- Development of guidelines for assessment and diagnosis required to ensure consistency and timely intervention

McConnell, 2021

Assessment of CVI

- Review article (2022): Consensus practice guide
- Concise Practice Guide by the Royal College of Ophthalmologists in the UK (2023)

Criteria for a diagnosis:

1. Presence of risk factor
2. Reported or observed atypical visual behaviour
3. Verifiable visual dysfunction on examination

- CVI may be present if 2 of the 3 criteria are met
- CVI highly likely to be present if all 3 criteria are met

Pilling et. al. 2022

Assessment of CVI: Presence of risk factors

Structured History-taking

Careful history to establish presence of risk factors:

- Pregnancy and birth history
  - Maternal infection
  - Prematurity with complications – resuscitation, hypoxic ischemic encephalopathy, white matter damage (periventricular leukomalacia)

- General health
  - Brain disorders
  - Meningitis/encephalitis
  - Chromosomal / genetic disorder
  - Epilepsy / infantile spasms
  - Metabolic disorders
  - Closed head injuries

Assessment of CVI: Reported / observed atypical behaviour

CVI questionnaires

- Visual skills Inventory for children – Ulster University
- Teach CVI screening lists 1-3
- Five questions

NB. CVI questionnaires should not generally be used for screening purposes: they yield too many false positives.[1]

1. Genderen et. al. 2012

CVI questionnaires – examples of questions

Multiple questions describing behaviors commonly associated with CVI, scored on a Likert scale (never, rarely, sometimes, often, always)

Ulster University Vision Resources
CVI Inventory Younger Child 4-6 years (2016)

	Never	Rarely	Sometimes	Often	Always	Not Applicable
Does your child…						
1. trip over toys and obstacles on the floor?	○	○	○	○	○	○
2. have difficulty walking down stairs?	○	○	○	○	○	○
3. trip at the edges of pavements going up?	○	○	○	○	○	○
4. trip at the edges of pavements going down?	○	○	○	○	○	○
5. appear to 'get stuck' at the top of a slide/ hill?	○	○	○	○	○	○

Abstract from the Ulster University CVI Inventory: Younger child 4-8 years (2016)

CVI behaviors frequently reported in clinic settings

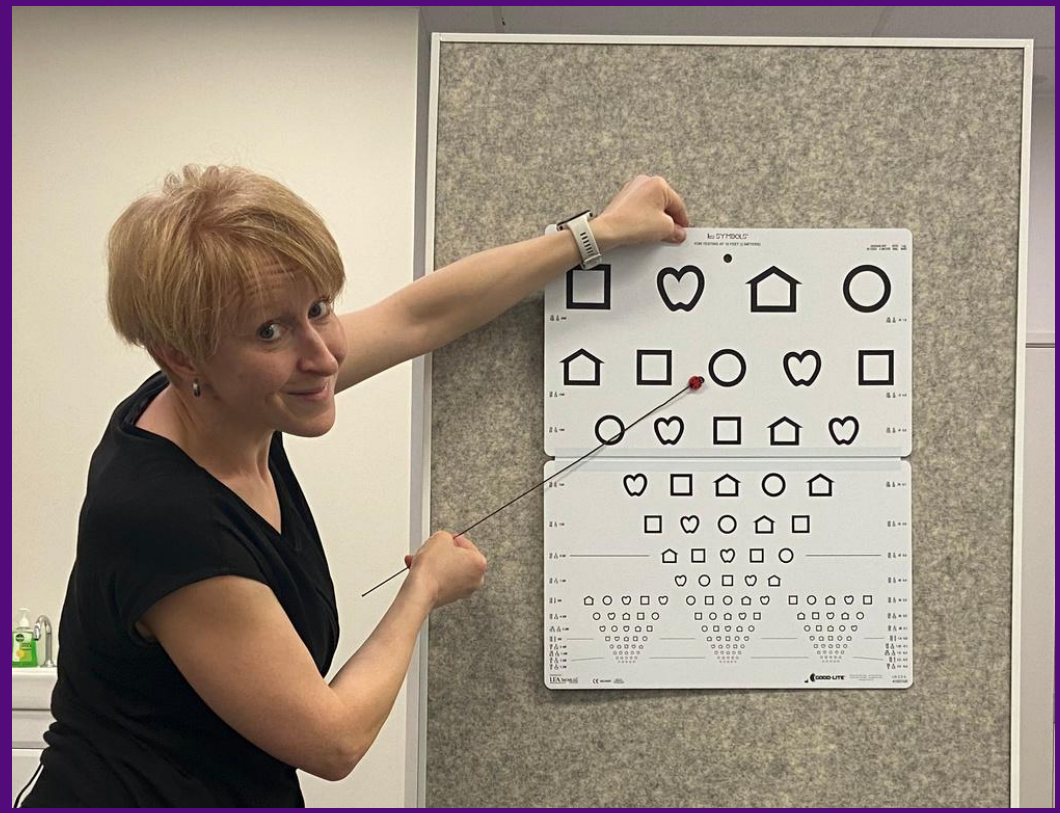
- Child cannot visually locate parent on pick-up from daycare
- Child will climb up to the slide, but won’t go down the slide, or will only go down head-first
- Child walks out in front of traffic

ASSESSMENT OF CVI: VERIFIABLE VISUAL DYSFUNCTION ON EXAMINATION

- Visual fields with Gross Peripheral Perimetry
- Looking for lower field defects or hemianopia

LEA SHAPES ACUITY CHART

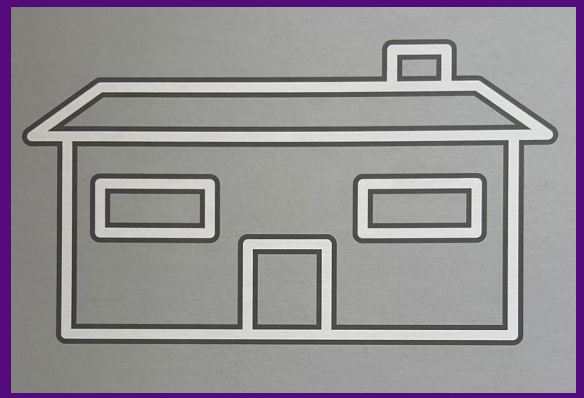
- GOLD STANDARD FOR CHILDREN
- Correlates with LogMAR letter chart
- Good repeatability
- Children can name / match shapes
- Adjustments: finding all the circles/houses



Visual Acuity

CARDIFF CARDS

- PREFERENTIAL LOOKING
- VANISHING OPTOTYPES
- Designed for younger children (18 months - 3 years) BUT great for children (and adults) with complex disabilities



Visual Acuity

LEA Paddles

- PREFERENTIAL LOOKING TEST
- Can be used on babies
- Good for children and adults with intellectual disability
- NOT comparable with letter/shape charts
- Adaptations: any stripes?

Visual Acuity

BRADFORD VISUAL FUNCTION BOX

- Selection of high contrast items
- Black and white books
- Light up toy
- Bright toys
- Beads presented in free-space

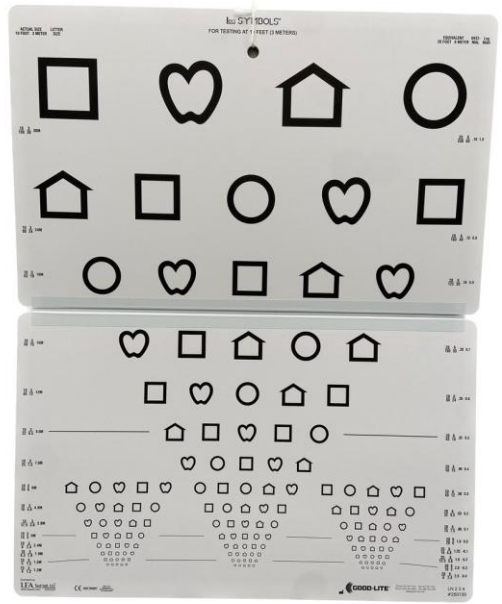
Visual Acuity

Pilling et. al. 2016

ASSESSMENT OF CVI: VERIFIABLE VISUAL DYSFUNCTION ON EXAMINATION

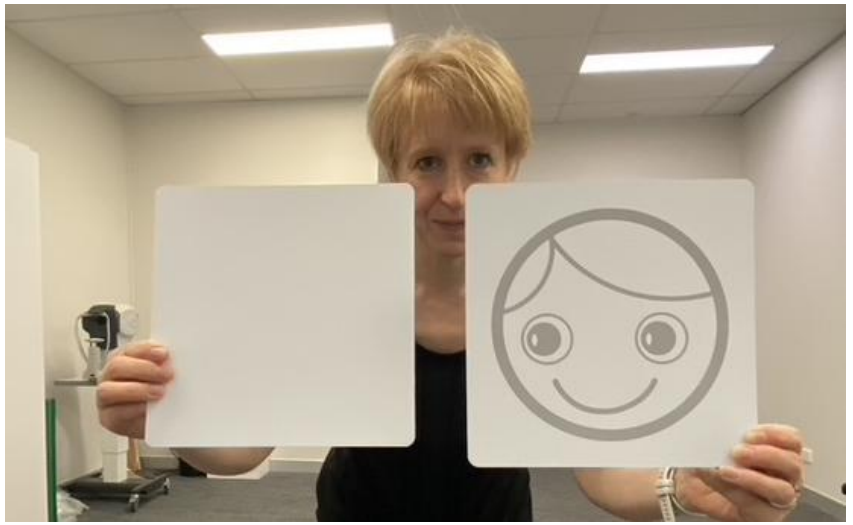
Observing <u>how</u> child is performing / interacting with the test.

- Visual latency
- More responsive when target held on one side
- Does noise override ‘seeing’?
- Size of letters seen when single, compared to ‘crowded’



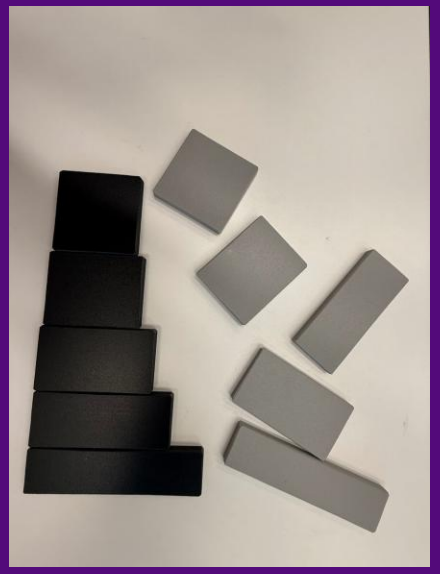
Assessment of CVI: Verifiable visual dysfunction on examination

- Contrast sensitivity



ASSESSMENT OF CVI: VERIFIABLE VISUAL DYSFUNCTION ON EXAMINATION

- Visually guided movement e.g. post a shape
- LEA puzzle: visual / tactile completion?
- Size perception: match shapes by size



ASSESSMENT OF CVI: VERIFIABLE VISUAL DYSFUNCTION ON EXAMINATION

- Ability to detect moving objects
- Detect a ‘new’ object introduced into the visual field
- Visually guided reach

WHY IS RECOGNITION IMPORTANT?

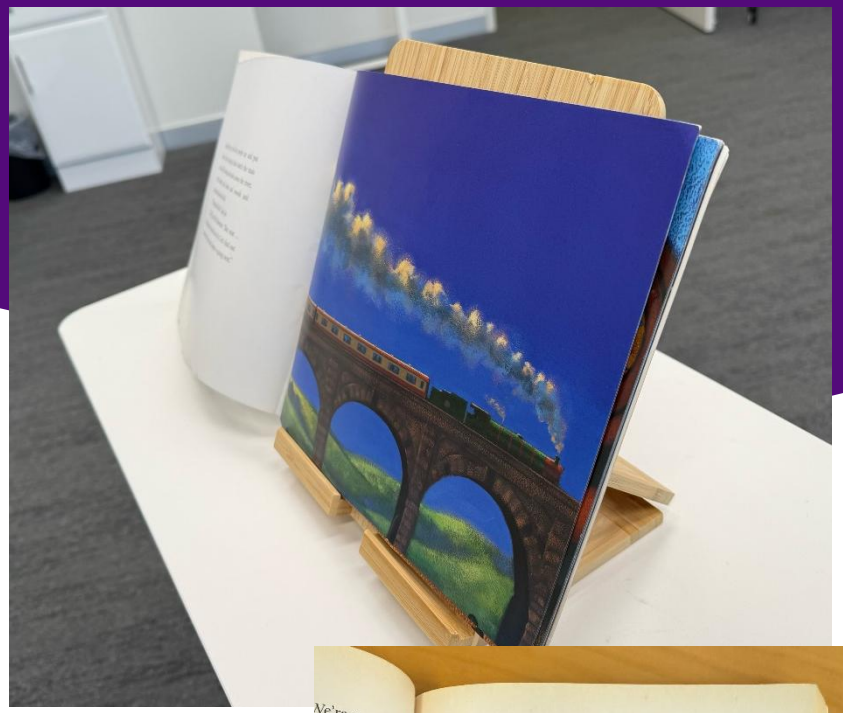
- No treatments
- Adjust environment to ensure full access
- Tailoring of therapies, based on knowledge of visual profile
- Without right adjustments, access to therapies and learning limited

WHAT MIGHT SUPPORT LOOK LIKE

Lower field defect

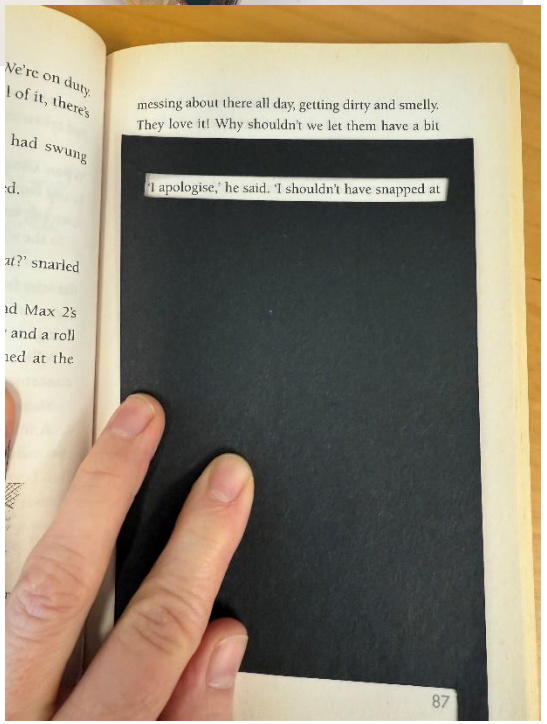
- Keep floors clear
- Keep furniture in the same place
- Verbal warnings when approaching stairs / kerbs
- Use a wheeled toy / stick to help child interpret where ground is
- Coloured markings on edges of steps
- Orientation and mobility training
- Practice on small slides first, with adult support
- Use a slope board to place learning material in upper field

WHAT MIGHT SUPPORT LOOK LIKE



Finding the next word / line when reading

- Using a finger to trace the lines
- Masking off additional print with plain card
- Use a typoscope
- Increase size and spacing of lines and words



WHAT MIGHT SUPPORT LOOK LIKE

Difficulties catching a ball

- Use a brightly coloured balloon - large and slow moving
- Balloon filled with rice - to give additional sound cues
- Try large, brightly coloured balls

SCREENING

CVI 5 questions:

Does your child:

1. Have difficulty walking down stairs?
2. Have difficulty seeing things that are moving quickly, such as small animals?
3. Have difficulty seeing something that in pointed out in the distance?
4. Have difficulty locating an item of clothing in a pile of clothes?
5. Find copying words or drawings time consuming and difficult?

≥2 warrants consideration for CVI

Dutton; 2019

SUMMARY

- CVI is common: 1 in every classroom
- CVI is under-diagnosed: 1 in 5
- Children with undiagnosed CVI experience problems with learning, friendship, and high levels of anxiety and emotional issues
- A recognised, multidisciplinary diagnostic pathway is needed in Australia
- Strategies are available for supporting children with CVI, regardless of levels, but we have to find them first!

THANK YOU FOR LISTENING.
ANY QUESTIONS?

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