

Concise Practice Points



The Role of the Eye Clinic in Clinical Assessment, Investigation, Diagnosis, and Initial Management of Paediatric Cerebral Visual Impairment

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Introduction

Concise Practice Points are carefully developed advice on individual elements or a combination of elements of patient care to help inform clinicians to make better decisions. A Concise Practice Point offers clarity for clinicians in subject matter which is frequently encountered but where there are areas of uncertainty or lacking in robust evidence. As such, the diagnosis of cerebral visual impairment is a suitable topic.

What history, examination and investigations should be undertaken for a child attending an NHS paediatric ophthalmology clinic when referred with suspicion of cerebral visual impairment?

What findings would constitute a diagnosis of cerebral visual impairment?

What should be written in a report to the parents, healthcare professionals and wider support team?

What support/advice/signposting/onward referral should be offered by a general paediatric ophthalmology team?

The CVI CPP scope was focused on the following review questions drawn from round-table discussions with orthoptists, optometrists, ophthalmologists, parents, teachers of the visually impaired and third sector organisations. It represents current understanding of the topic and acknowledges the evolving nature of practice. It will be regularly reviewed in the light of emerging evidence.

Methods

PubMed and Cochrane Library databases were systematically searched in March 2021 using defined search terms. Articles were included if they:

- Were research papers, conference abstracts or research protocols in peer-reviewed scientific journals or text books
- Included a clinical investigation of CVI in children

Reference lists included articles were also reviewed to locate other articles which may not have been identified from the initial search.

Articles were excluded if they were not published in English language or were single case reports.

Full details of the review have been published in Eye¹.

In addition a focus group of qualified teachers of the visual impaired was undertaken to provide advice around the information which should be included in a written report to education.

A draft version of the CPP was reviewed by members of the UK CVI special interest group (eye health professionals from the UK, primarily orthoptist and paediatric ophthalmologists) for their input and suggestions.

Cerebral Visual Impairment is the commonest cause of visual impairment in children in the developed nations²⁻⁵. It is a common condition which every ophthalmologist may expect to encounter in a paediatric ophthalmology clinic.

Cerebral Visual Impairment is defined as a verifiable visual dysfunction which cannot be attributed to disorders of the anterior visual pathways or any potentially co-occurring ocular impairment⁶. CVI is an umbrella term used to describe problems with vision relating to the brain, which include abnormalities in visual acuity, or contrast sensitivity or colour; ocular motility; visual field and the processing of visual input. CVI can occur in isolation, or in combination with ocular causes of visual impairment.

Children with Cerebral Visual Impairment may have significant visual difficulties affecting daily life, education, and independence. The spectrum of dysfunction and presentations requires a curious, proactive, and flexible approach to history taking, examination and diagnosis which will differ for each child.

There are three main elements to the diagnosis of cerebral visual impairment: where there is a positive finding in all three areas, it is probable that CVI is present. Where there is a positive finding in 2 of the 3 elements from Box 1 below, CVI should be considered, and further evidence to support or exclude a diagnosis sought from parents or other professionals.

Box 1: Cerebral Visual Impairment Diagnostic Elements

1	Developmental anomaly / positive risk factor for cerebral visual impairment	e.g., Down's syndrome, ex premature, cerebral palsy, hypoxic ischaemic encephalopathy, hydrocephalus, developmental impairment / intellectual disability
2	Externally observed or reported symptoms of cerebral visual impairment / atypical visual function	e.g., Unexplained failure to progress at school; difficulty finding objects; difficulty with complexity; clumsiness; difficulty recognising people, shapes, places; difficulty with seeing moving objects; eccentric visual behaviours; may include information from inventories / questionnaires
3	CVI-related visual dysfunction elicited on examination in any combination of areas below (Table 1)	e.g., Traditional acuity tests, crowding test, contrast sensitivity; confrontation field, dynamic retinoscopy, refraction, oculomotor function (pursuits, saccades), simple clinic based perceptual tests

Table 1: Commonly encountered Cerebral Visual Impairment related visual dysfunctions

Lower Visual Function Impairment	Higher Visual Function Impairment Ventral Stream	Higher Visual Function Impairment Dorsal Stream
<p>Reduced binocular visual acuity (in particular with crowded testing) not due to refractive error</p> <p>Abnormal fixation</p> <p>Visual field deficit / inattention</p> <p>Oculomotor impairment (jerky smooth pursuit, inaccurate saccades)</p>	<p>Visual memory / recognition (impaired ability to name shapes, objects, letters, recognise faces, or facial gesture)</p> <p>Route finding / orientation (impaired ability to 'map' their world or a room to orient effectively through it) and to judge visual space</p>	<p>Binocular Crowding ratio >2.0 (Disparity in visual acuity measurement with crowded vs uncrowded optotypes)</p> <p>Impaired ability to notice more than one object at a time (divided visual attention)</p> <p>Difficulty discerning individual elements from a background scene (simultagnosia)</p> <p>Movement perception (impaired ability to see moving objects OR impaired ability to see objects unless they are moving)</p> <p>Visual guided movement (impaired ability to reach for / locate objects / navigate steps / kerbs)</p>

It is the role of the ophthalmologist and eye care team:

- To exclude and / or manage co-existing ocular conditions (e.g., optic disc anomalies, structural anomalies such as cataract, coloboma), refractive error and accommodation dysfunction
- To establish by history taking risk factors for, and symptoms of, CVI to enable proactive detection of visual dysfunction or behaviours indicative of such
- To integrate eye clinic findings in conjunction with paediatric and developmental / disability teams, viewing visual dysfunction in the context of any associated neurodevelopmental or other medical conditions, taking into account the differential diagnosis of symptoms of CVI

*Diagnosis should **not** be delayed while awaiting the child's development to allow for formal acuity, fields, or perceptual testing.*

Clinical inquiry requires a flexible approach, individualised for the child. Suggested tools for children with a range of developmental and cognitive abilities are listed in Appendix 1. Structured questions relating to the child's visual function can be helpful in eliciting signs and symptoms of cerebral visual impairment and may supplement general history taking⁷⁻¹⁸ (Examples are listed in appendix 2). Whilst not essential for diagnosis, they can be useful in devising an examination strategy for the individual child.

For the minority of children who are found by the eye care team to have features of CVI but have not been previously diagnosed with a neurodevelopmental anomaly, it is essential that a paediatric assessment is arranged to identify any additional developmental conditions which may co-exist. The referral should specify that CVI is suspected and ask for a neurodevelopmental perspective to help establish whether there is a specific visual impairment (ie CVI) or a more global delay in development that explains the behaviours causing concern. In some children both CVI and a neurodevelopmental condition may co-exist and multidisciplinary working is essential to ensure all diagnoses and associated needs are recognised.

Costly investigations such as neuroimaging^{4 19 20 21} or electrodiagnostic testing^{21 22} are often not required in the diagnosis of cerebral visual impairment. They can be useful when there is diagnostic uncertainty or in the presence of severe developmental impairment / intellectual disability to identify coexisting ocular / neurological pathology particularly occult retinal disease.

Visuoperceptual testing does not form part of the core visual assessment; however, it is important that ophthalmologists arrange or suggest referral to professionals (e.g., neurodisability team (includes paediatrician and OT), psychologists (neuro or educational) and QTVIs) in the local area, where testing may contribute to a broader understanding of the areas of a child's visual dysfunction and the assessor has the competencies to carry out and interpret more detailed visuoperceptual assessment.

Some children with normal acuity and field will require such testing in order to demonstrate verifiable visual dysfunction to fulfil the diagnostic criteria and receive a confirmed diagnosis of cerebral visual impairment ^{4 19 23-28}.

It is essential to explain to parents that while there is no 'cure' for Cerebral Visual Impairment, it is not true to say that 'nothing can be done'; neither is it helpful to state simply 'there's nothing wrong with his / her eyes'. In some cases, improvements in function can be seen over time, and support from education, habilitation, OT and family can be transformative, as children learn strategies to maximise the efficiency of their vision, finding ways to 'make it easier to see' ^{9 29}(appendix 3).

Examples of diagnostic pathways are shown in appendix 5. Whilst it is beyond the scope of this document to specify a model of care. It is expected that where CVI is identified, or suspected, ophthalmologists and their teams undertake the following steps:

- Acknowledge the presence or possibility of CVI and provide an explanation of the nature of the child's visual disability, including signposting information
- Manage refractive error including bifocals for children with poor accommodative function
- Offer information on initial strategies based on clinical findings (e.g., field of attention, difficulties with tracking moving objects, difficulty with clutter). If there is uncertainty about the diagnosis pending further assessment, strategies can be offered as a "trial" which provides useful information either way, about how helpful the strategies are for that child
- Communicate with the local Neurodevelopmental Paediatric service about the concerns, to involve them in the possible diagnostic formulation and discuss what further paediatric assessment is optimal or can be offered
- Contribute to effective report writing encompassing the child's visual function, beyond a simple acuity measurement. Sharing this report with parents and the wider team supporting the child.
- Arrange referral to education, social care and voluntary services (sensory support team, qualified teachers of the visually impaired, habilitation services)
- To consider sight impairment registration, (or notification to VINCYP if in Scotland) regardless of visual acuity, if the child is judged to be significantly disadvantaged by their visual function.
- Refer to Eye Clinic Liaison Officer (where available) and signpost charity / support organisations (appendix 4)
- Where appropriate, to arrange onward referral for further assessment and support (e.g. neurodisability team, sensory services, occupational therapy, psychology, low vision clinic)

Appendices

Appendix 1: Examples of visual assessment tools for children with and without verbal / motor skills

Appendix 2: Cerebral Visual Impairment Inventories / Questionnaires

Appendix 3: Strategies to support children with CVI

Appendix 4: UK CVI charities and patient support organisation

Appendix 1: Examples of visual assessment tools for children with and without verbal / motor skills

Visual Function	Test for verbal / motor children	Test for pre-verbal / pre-motor children
Visual acuity (Near and Distance)	log MAR, Kay pictures	Cardiff acuity test Teller acuity test Questions 1-11 of PreViAs ¹⁷
Visual attention – Duration distance and object size	By observation	Bradford Visual Function Box ^{5 30} Mirror test ^{13 31} Puppet Face Neonatal Assessment Visual European Grid ¹³
Crowding ratio ³² ³³ for Near and Distance (both eyes open)	log MAR, Kay pictures	
Contrast Sensitivity or Low Contrast Acuity	Hiding Heidi ³⁴ Low contrast letter charts (Lea, ETDRS)	Hiding Heidi Cardiff Contrast sensitivity test ³⁵
Confrontation visual fields, including inferior visual field	Using appropriate object size related to visual acuity / attention	Puppet Face
Divided visual attention	Ability to detect a 'new' object in a different areas of visual field	Ability to detect a 'new' object in a different areas of visual field
Pupil reactions		
Pursuit and saccades	Using appropriate object size and mindful of visual field inattention: horizontal and vertical	Using appropriate object size and mindful of visual field inattention: horizontal and vertical
Accommodation	Dynamic retinoscopy	Dynamic retinoscopy
Object orientation	Lea mailbox ³⁶ (for children over developmental age 4 years)	
Shape identification	Lea rectangles ^{36 37} (for children over developmental age 6 years)	Lea 3D puzzle test
Oculo-motor coordination (Reach&Grab)	Using a suitable size object based on visual acuity. Observing accuracy, speed, and eye / head alignment during task	Using a suitable size object based on visual acuity. Observing accuracy, speed and eye / head alignment during task
Ability to detect moving objects	CVIT 3-6 ^{38 39}	By observation

Appendix 2: Cerebral Visual Impairment Inventories / Questionnaires

Tool	Notes / reference
Insight / Visual Skills Inventory	8 10 www.ctevh.org/Conf2017/Workshops/407/407%20-%20Insight.html 52 item questionnaire with links to strategies
PreVias	17 Tool for children under 12 months
TeACH	www.teachcvi.net/ 3 questionnaires for children of differing cognitive abilities 15
Flemish CVI questionnaire	16 https://cdn-links.lww.com/permalink/coop/a/coop_2012_05_31_lehman_656_sdc1.pdf 46 item tool designed to draw out behaviours associated with CV

Appendix 3: Strategies to support children with CVI

Insight: Approaches for Visual Perceptual Difficulties⁹

www.ctevh.org/Conf2017/Workshops/407/407%20-%20Insight.html

Make It Easier To See²⁹

www.makeiteasiertosee.co.uk

Ulster University Cerebral Visual Impairment Strategies

www.ulster.ac.uk/research/topic/biomedical-sciences/research/optometry-and-vision-science/vision-resources/professionals/cerebral-visual-impairment-assessment

Colour Tents⁴⁰

www.cvisociety.org.uk/news.php?cat_id=250

Appendix 4: UK CVI charities and patient support organisations

www.cvisociety.org

www.cviscotland.org

Appendix 5

Cerebral Visual Impairment (CVI) Diagnostic Pathway, Visual Impairment Network for Children and Young People, NHS Scotland⁴¹

Cerebral Visual Impairment and Clinical Assessment: The European Perspective⁴²

The multidisciplinary guidelines for diagnosis and referral in Cerebral Visual Impairment (CVI) (Netherlands)⁴³

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