Vision in children with Cerebral Palsy

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Introduction

• Many children with cerebral palsy have visual disorders (10% severe visual impairment)
• May be due to cerebral problem (cerebral visual impairment, visual field defect), or ocular disorder (refractive, amblyopia, squint, eye abnormality)
• Important need to assess visual deficits, and manage associated conditions to maximise visual potential and provide appropriate support (education, rehabilitation)
Vision in children with Cerebral Palsy: overview

- Visual development and maturation
- Refractive error
- Amblyopia
- Strabismus, nystagmus
- Delayed visual maturation
- Cerebral visual impairment
- Prematurity-ROP, PVL, IVH
- Neonatal encephalopathy

- Assessment of vision in children with CP
- Management of children with VI and CP
Visual development and maturation

- Visual development from 6 weeks – fixes and follows readily by age 2 months
- Binocular function develops in first 6 months
- Amblyopia and strabismus are common in children with cerebral palsy
- Amblyopia managed with glasses/patching/atropine up to the age of 8-9 years
Strabismus in CP

- 56% of patients with CP had strabismus
- Exotropia common
- Large angle esotropia
- Rarely accommodative
- Amblyopia common
- Often nystagmus
- Surgery when indicated (discussion with parents/child)-rarely binocular potential


*Strabismus in children with cerebral palsy.*

Erkkilä H¹, Lindberg L, Kallio AK
Refractive error

• Usually hypermetropia in infancy
• Up to 3 dioptres hypermetropia does not need correction if no strabismus
• Need glasses (to prevent amblyopia) if
  – Over 3 dioptres of myopia
  – Astigmatism > or equal to 2 dioptres
  – Anisometropia (>1 dioptre)
• Ametropic amblyopia if uncorrected refractive error
• Relation to squint- partially accommodative esotropia
• Myopia in ex-prematurity
Nystagmus in CP

- Common
- Maybe due to intracranial abnormality, prematurity
- No oscillopsia in congenital nystagmus
- Face turn to utilise null zone, may be variable
- Commonly horizontal, maybe vertical or multiplanar
- ERG done to rule out other cause
Visual field defects in CP

- Common in CP
- Congenital hemianopia (occipital cortex defects, periventricular leukomalacia, periventricular haemorrhage)
- May have little effect on functioning when congenital (may not be apparent until older)
- May be difficult to assess
- Face turn to defective field on fixation
- May have exotropia
- May have corresponding optic atrophy

Left occipital cortex involvement
Co-existent eye disease and CP

- Optic nerve hypoplasia (schizencephaly, septo-optic dysplasia)
- Optic atrophy (trans-synaptic degeneration, hydrocephalus, PVH)
- Retinopathy (Joubert, Zellwegers)
- Retinal abnormality (Aicardi)
- Cataracts (Nance-Horan, Micro syndrome)
Delayed visual maturation

• Occurs when delay to normal visual milestones
• May occur with normal eye and systemic exam (type 1)
• In the presence of neurological impairment/ developmental delay, seizures (type 2)
• May occur in presence of other ocular condition eg albinism, optic nerve hypoplasia (type 3)
• May be difficult to distinguish between DVM and CVI initially
• Gradually improve between 6-24 months
• Maybe eventually normal visual acuity and function
Cerebral visual impairment in CP

- Commonest cause of all visual impairment in developed countries
- Can effect visual acuity, contrast sensitivity, visual fields (lower field common), perceptual and cognitive visual impairment
- Strabismus common
- Light gazing and photophobia can both occur in severe CVI
- Normal ocular examination
Cerebral visual impairment - perceptual visual dysfunction

- Wide range of visual impairment and dysfunction
- May be difficult to quantify when coexistent motor or intellectual dysfunction make assessment difficult
- Unique problems for each individual
- Visual inattention and impaired visual search (locating toys on patterned background, seeing things at a distance)
- Impaired location in 3-D - difficulty in getting around
- Impaired recognition (people, shape, objects)
- Variable function- depends on attention, visual overload, fatigue
Cerebral visual Impairment- dorsal and ventral stream visual processing

Dorsal stream
(occipital and posterior parietal cortex)

Ventral stream (occipital and inferotemporal cortex)
The perceptual visual system-dorsal stream

• Dorsal stream and posterior parietal cortex assimilate incoming visual information to bring about immediate guidance of skilled actions (facilitates accurate guidance of movement of the body)

• Damage causes inaccurate visual guidance of movement (despite conscious visuospatial awareness)
The perceptual visual system-ventral stream

- Ventral stream and inferotemporal cortices
- Store previous visual experiences and serve conscious recognition and understanding of what is seen
- Damage impairs visual recognition (faces, objects and shapes) and route finding
- May use voices and other cues
- Visual memory may be impaired-copying, drawing, remembering where things are
Questions to assess CVI

• Does your child have difficulty walking downstairs?
• Does your child have difficulty seeing things which are moving quickly?
• Does your child have difficulty in seeing something which is pointed out in the distance?
• Does your child have difficulty locating an item of clothing in a pile of clothes?
• Does your child find copying words or drawings time-consuming and difficult?
Assessment of CVI

• 51 item inventory to assess 7 areas-
• Visual field or localised attention
• Perception of movement
• Visually guided movements of the body
• Visual attention
• Coping with crowded scenes or environments
• Recognition and orientation

From: Gordon Dutton and Richard Bowman
in Paediatric Ophthalmology and Strabismus 4th edition 2013
Management of the child with CVI

- Compensation or adaptation to the impairment
- Environmental modifications or the use of devices
- Training of the impaired functions

- Reduced acuity/colour vision- enlarge text, double space, good contrast
- Visual field defects-trace text with finger/ruler, seat position in classroom, careful guidance around new area
- Locating items- organized storage, avoid clutter, labelling
- Finding food on plate- avoid patterns, separate portions
Prematurity: retinopathy of prematurity

- CP often related to prematurity
- Babies ages <32 weeks and <1500g screened for ROP in UK
- Relation of ROP presence and severity to high oxygen levels
- Approximately 6-8% of babies less than 1251g will require treatment for ROP (laser treatment or anti-VEGF injections)
- High myopia
- Visual impairment from retinal detachment or macular dragging (or non-ocular causes)
Retinopathy of prematurity

- Zones 1-3 of vascular development
- Stage 1-3 of ROP
- Treated with diode laser ablation of avascular retina
Periventricular leukomalacia

- Associated with prematurity and CP
- Is a major cause of visual disability in many parts of the world
- May be associated with retinopathy of prematurity
- Variable visual impairment- severe to minor visual field defects (usually inferior)
- Tonic downward gaze in infants (dolls head manoeuvre- not an upgaze palsy)-usually resolves
Periventricular leucomalacia

- Often large esotropia (more common than exotropia)
- May be manifest or latent nystagmus
- May be reduced accommodation
- Normal sized optic nerve with enlarged cup (pseudo-glaucoma)
Intraventricular haemorrhage

- Ocular pathway involvement-visual field defects
- Raised Intracranial pressure and hydrocephalus may lead to optic atrophy
- Causes 16% of congenital optic atrophy

Childhood optic atrophy (n=272).
Mudgil AV, Repka MX.
Visual impairment in neonatal encephalopathy

- Involvement of occipital cortex
- Variable extent of visual loss - complete blindness to small VF defect
- Variable visual performance - best in familiar surroundings, detection of motion or colour, isolated letters rather than whole words
- Usually normal ocular exam (normal pupils, no nystagmus)
- May have supranuclear ocular motor abnormalities (horizontal gaze palsies, smooth pursuit difficulties)
- May have optic atrophy
- VEPs may be helpful
- Usually improvement over time
Ophthalmic assessment of child with CP

• History-pregnancy, birth history, medical history and medications
• Visual development
• Nystagmus, squint
• Cerebral Visual impairment questionnaire
Vision assessment in child with CP

- Observation of visual behaviour
- Central, steady, maintained fixation each eye
- Ability to track small toy with each eye
- Ability to follow light/respond to light
- Snellen/LogMar near and distance acuity
- May need grating acuity- preferential looking cards, Cardiff cards (but overestimate acuity)
- Kays pictures
Ocular assessment in CP

• Colour vision-Ishihara, city- if possible
• Ocular movements- (ocular motor apraxia-impaired saccades, gaze palsies, impaired smooth pursuit)
• Fundoscopy-dilated (retinal problem, optic nerve)
• Refraction-cycloplegic (dynamic retinoscopy -accommodation problems common in CP)
• Visual field- to confrontation, Goldman, Humphrey, new technologies for VF testing in children
• Electrodiagnostics (ERG/VEP)
• (MR scanning)
Visual evoked potential

- Provides evidence of visual function
- Optic pathway integrity
- Pattern VEP down to 50’ or smaller check sizes suggests good visual function
- Flash VEP only suggests rudimenta
Electroretinography (ERG)

- Retinal response in light and dark adapted conditions distinguishes rod and cone dysfunction
- Photoreceptor or inner retinal dysfunction
- Rules out a retinopathy if nystagmus/poor vision
Management of the child with visual problems and CP

- School support- Visual Impairment Teachers
- Well known environment and peer group
- Identify problems and strategies for overcoming them

- Paediatric Ophthalmologist-
- Specialist clinics (multidisciplinary)
- Certificate of visual impairment
Summary: challenges in managing vision in children with Cerebral palsy

• Role of ophthalmologist/orthoptist is to assess visual defects as fully as possible—may be difficult in clinic
• Maximize visual potential/provide treatment
• Correct refractive error when needed
• Treat ambyopia when needed
• Strabismus surgery when indicated
• Counselling, advice and support for parents/teachers/carers