Title: Vision Problems in Children with Cerebral Palsy: Practical Strategies for Screening/identification, Referral, Diagnosis and Treatment
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Why do vision screening:
Eliminate preventable vision loss (amblyopia)

Theory behind vision screening:
Detect risk factors or causes of amblyopia with history and physical screening method (recognition acuity or instrument based)

Definition of amblyopia:
Amblyopia is decreased vision in one or both eyes due to abnormal development of vision in infancy or childhood. In amblyopia, there may not be an obvious problem of the eye. Vision loss occurs because nerve pathways between the brain and the eye aren't properly stimulated.
https://aapos.org/glossary/amblyopia

Causes of amblyopia:
- Uncorrected unilateral or bilateral refractive error
  - May be most challenging to discover with screening in uncooperative child or child with little ability to participate in typical letter or picture recognition based screening
- Strabismus
- Media opacity
- External structure blocking visual axis (ptosis, etc)

Latest AAP vision screening policy:
Donahue, Sean, Nixon, Cynthia. Visual System Assessment in Infants, Children, and Young Adults by Pediatricians. *Pediatrics*. 2016;137(1):28-30. doi:10.1542/peds.2015-3596

Latest AAP vision screening technical report: Donahue, Sean P., Baker, Cynthia N. Procedures for the evaluation of the visual system by pediatricians.(Report). *Pediatrics*. 2016;137(1):e20153597-e20153597. doi:10.1542/peds.2015-3597

Current Recommendations

- Instrument-based screening (photoscreening) is recommended for children 12-months of age and older unless they can reliably perform visual acuity testing with eye charts.
- Direct measurement of visual acuity using eye charts remains the gold standard for vision screening and can often begin by 4-years of age.

TABLE 1 Periodicity Schedule for Visual System Assessment in Infants, Children, and Young Adults

Assessment	Newborn to 6 mo	6–12 mo	1–3 y	4–5 y	6 y and older
Ocular history	x	x	x	x	x
External inspection of lids and eyes	x	x	x	x	x
Red reflex testing	x	x	x	x	x
Pupil examination	x	x	x	x	x
Ocular motility assessment	—	x	x	x	x
Instrument-based screening[a] when available	—	[b]	x	x	[c]
Visual acuity fixate and follow response	x[f]	x	x	—	—
Visual acuity age- appropriate optotype[d] assessment	—	—	x[e]	x	x

[a] *Current Procedural Terminology* code 99174.

[b] The American Academy of Ophthalmology (AAO) has recommended instrument-based screening at age 6 mo. However, the rate of false-positive results is high for this age group, and the likelihood of ophthalmic intervention is low.[16] A future AAO policy statement will likely reconcile what appears to be a discrepancy.

[c] Instrument-based screening at any age is suggested if unable to test visual acuity monocularly with age-appropriate optotypes.

[d] *Current Procedural Terminology* code 99173.

[e] Visual acuity screening may be attempted in cooperative 3-y-old children.

[f] Development of fixating on and following a target should occur by 6 months of age; children who do not meet this milestone should be referred.

Pediatrics. 2016;137(1):28-30. doi:10.1542/peds.2015-3596

Examination of the eyes and visual system should begin in the nursery and continue throughout both childhood and adolescence during routine well-child visits in the medical home.
http://pediatrics.aappublications.org/content/early/2015/12/07/peds.2015-3596

Children with these conditions should be referred for a complete eye examination:

- Premature infants as per AAP/AAO/AAPOS guidelines
- Congenital cataracts
- Retinoblastoma
- Metabolic or genetic diseases
- Significant developmental delay or neurologic difficulties
- Certain systemic diseases
- Family history of ocular disease

Special Populations with developmental delay

- Down Syndrome
- Neurofibromatosis
- Populations at risk for cortical visual impairment
  - Cerebral palsy
  - Hydrocephalus
  - Seizures
  - Neonatal encephalopathy
  - Neonatal hypoglycemia
  - Brain tumor
  - Traumatic brain injury
  - Stroke
  - Neonatal systemic infection (Zika, HSV, etc)

Referral criteria concerning inability to perform screening
Any child who cannot be adequately screened in the medical home should be referred to an eye care professional experienced in the care of children.

Pearls for screening children with developmental delay

- Perform modified screening/examination consistent with developmental age
  - Recognition acuity
    - Fix and follow
    - Have some interesting toys
    - Symbols or letters
    - Matching is a useful technique
  - Instrument based screening
    - Quick
    - Noncontact
    - Use distraction techniques

Instrument based vision screening

- Requires very little cooperation from child
- May detect abnormalities other than refractive error
- Produces data for chart documenting performance of test and results
- Some methods may provide interpretation

Photoscreeners

- Uses optical quality of red reflex to identify amblyopia risk factors such as refractive error as well as:
  - ocular alignment (strabismus)
  - media opacities (cataract)
  - abnormalities of periocular structures (ptosis)
- Tests binocularly
- Data interpretation:
  - By operator
  - Central reading center
  - computer

Autorefractors

- Handheld device much more useful in children
- Involves optics for determination of refractive error
- Sensitivity of 81% and specificity of 88%
- Some are monocular and some are now binocular
- Monocular units will not detect strabismus
- Data interpretation
  - Numeric results
  - Must be analyzed by operator or device itself

Barriers to adoption of instrument based screeners by primary care

- Costs
  - Direct: costs of device and supplies
  - Indirect: staff time for training and maintaining competency, provider time for interpretation
- Reimbursement
  - CPT code: 99174 with RVU 0.69
  - No guarantee that code will generate payment

Instrument based vision screening pearls

- Early age appropriate screening should occur in the medical home
- May be performed in children 12 months to 3 years of age
- Alternative for older children who cannot cooperate for recognition acuity rtesting
- In children ages 3 to 5 years of age, recognition visual acuity and standard examination techniques (corneal light reflex, examination of red reflex, etc) remains a useful and viable technique

A Practical Guide for Primary Care Physicians: Instrument Based Vision Screening In Children
www.ChildrensEyeFoundation.org/SEE

Systemic comorbidities in children with cerebral palsy:

- Retinopathy of prematurity
- Seizure disorders

Ocular conditions seen in children with cerebral palsy:

- Strabismus
- Amblyopia
- Refractive errors
- Nystagmus
- Saccade and pursuit deficiency
- Optic atrophy
- Cortical/cerebral visual impairment

Black, P D. Ocular defects in children with cerebral palsy. *British Medical Journal*. 1980;281(6238):487-488. doi:10.1136/bmj.281.6238.487
Katoch, Sabita, Devi, Anjana, Kulkarni, Prajakta. Ocular defects in cerebral palsy. *Indian Journal of Ophthalmology*. 2007;55(2):154-156. doi:10.4103/0301-4738.30717

Strabismus
Strabismus or misaligned eye can be seen in up to 50% of children who have cerebral palsy.

Treatment
Medical therapy

- Correct significant refractive error with glasses.
- Surgical intervention
- Patching of preferred eye or atropine penalization to prevent/treat amblyopia (poor vision due brain shutting off to eye not being used).
- Observation

Surgical

- Eye muscle surgery
- Botox injection of the eye muscle

Esotropia more common than exotropia in children with cerebral palsy.

Overcorrection is more common after eye muscle surgery in children with cerebral palsy.
Some studies show variability of alignment in children who have cerebral palsy.

Delay in surgical intervention may contribute to worse surgical outcome.

It is reasonable to consider surgical intervention in children with cerebral palsy to prevent amblyopia, restore binocularity and improve social interaction and self image. Every case should be treated individually by the pediatric ophthalmologist and family after a discussion of risks and benefits.

Mary Louise Z. Collins (2014) Strabismus in Cerebral Palsy: When and why to Operate, American Orthoptic Journal, 64:1, 17-20, DOI: 10.3368/aoj.64.1.17

F. Miller etal (eds.), Cerebral Palsy, Springer International Publishing 2018; Strabismus Management in the Child with Cerebral Palsy; https:/doi.org/10.1007/978-3-319-50592-3_77-1

Refractive error
High refractive errors are more common in children with cerebral palsy.
Premature infants have an increased risk of high refractive errors.
Genetic factors may also contribute.
Significant refractive errors should be corrected.
Poor accommodation (focusing to overcome refractive error) may be deficient in children with cerebral palsy and require correction.

McClelland JF (2006) Accommodative dysfunction in children with cerebral palsy. Invest Oph Vis Sci 47:1824-1830

Saunders, Kathryn J, Little, Julie-Anne, Mcclelland, Julie F, Jackson, A Jonathan. Profile of refractive errors in cerebral palsy: impact of severity of motor impairment (GMFCS) and CP subtype on refractive outcome. *Investigative ophthalmology & visual science*. 2010;51(6):2885-2890. doi:10.1167/iovs.09-4670

Cortical/Cerebral Visual Impairment

Definition

- Bilateral visual impairment due to brain damage of the posterior visual pathway
- Eye structure is typically normal or the pathology found (optic atrophy) does not explain visual impairment
- Children with CVI display characteristic behaviors

Causes

- Structural: brain malformations, tumors
- Vascular: periventricular leukomalacia (PVL) secondary to prematurity, hypoxic/ischemic event, perinatal stroke
- Infectious: meningitis, encephalitis
- Inflammatory: vasculitis (inflammation of blood vessels)
- Trauma: TBI, nonaccidental trauma
- Metabolic: neonatal hypoglycemia, mitochondrial disease, lysosomal disorders
- Neurologic disease: seizure, hydrocephalus

Lueck AH, Dutton GN, editors. Vision and the Brain: Understanding Cerebral Visual Impairment in Children. New York, NY: AFB Press; 2015

Characteristics in infants or children with significant neurologic impairment

- Variable, poor or atypical response to visual stimuli
- Poor or inefficient use of visually guided (eg.: eccentric viewing or frequent looking away)
- Latency (delay in visual response)
- Difficulty with distance viewing
- Difficulty with novelty
- Preference for familiarity
- Light gazing (preference for looking at light)
- Color preference
- Difficulty with complexity
- Preference for certain visual fields
- Better visual performance with movement

Roman-Lantzy C. Cortical Visual Impairment: An Approach to Assessment and Intervention (second edition) New York, NY: AFB Press; 2018

Damage to connections of visual centers
Dorsal stream function – getting there

- Connections between occipital area to parietal areas
- Responsible for:
    - Finding objects in space
    - Figure/background
    - Extremity movement

- Examples of deficits:
    - Difficulty with steps or changes in surfaces
    - Inaccurate reach
    - Difficulty with complexity

Damage to connectors of visual centers
Ventral stream function – who or what is there

- Connect between occipital area and temporal lobe
- Responsible for:
    - Form recognition
    - Visual memory
- Examples of deficits:
    - Forget location of objects
    - Difficulty with recognition of faces, shapes, objects

Merabet, Lotfi B., Mayer, D. Luisa, Bauer, Corinna M., Wright, Darick, Kran, Barry S. Disentangling How the Brain is "Wired" in Cortical (Cerebral) Visual Impairment. *Seminars in Pediatric Neurology*. 2017;24(2):83-91. doi:10.1016/j.spen.2017.04.005

Bauer, Corinna M., Merabet, Lotfi B. Perspectives on Cerebral Visual Impairment. *Seminars in Pediatric Neurology*. 2019. doi:10.1016/j.spen.2019.05.001

Cortical vs. Cerebral Visual Impairment

- Various interpretations:
    - Outside the United States, the term cerebral is more commonly used
    - Some use terms interchangeably
    - Some use cerebral as a broader term which includes vision issues due to brain damage of more superficial parts of the brain (cortical) as well as deeper structures in the brain
    - Some use term cerebral to mean higher level perceptual and processing issues
    - Others use cortical as a subset of cerebral displaying specific characteristics

Recovery of visual function in CVI

- Most children have some degree of improvement
- Recovery can occur over months to years
- Degree of recovery cannot be predicted from imaging studies

*Matsuba CA, Jan JE. Dev Med Child Neurol. Long-term outcome of children with cortical visual impairment; 2006 Jun;48(6):508-12.
Watson T[1], Orel-Bixler D, Haegerstrom-Portnoy G; Optom Vis Sci.; Longitudinal quantitative assessment of vision function in children with cortical visual impairment 2007 Jun;84(6):471-80.

CVI Workup/Treatment

- Ophthalmologic evaluation with pediatric ophthalmologist
    - Perform history and physical
    - Treat ophthalmologic problems
    - Provide diagnosis of cortical visual impairment
    - Provide medical necessity necessary to obtain vision services
    - Refer for vision services
    - Provide educational information for family
    - Make specific recommendations for child based on characteristics
    - Provide information concerning child medical condition to family and team

CVI Workup/Treatment

- Functional evaluation performed by teacher of the visually impaired experienced in cortical visual impairment
    - History
    - Functional evaluation
    - Recommendations based on characteristics
    - Periodic reassessments

Multidisciplinary team approach: Effect of CVI on performance should be considered in all aspects of ADL

- parents/family/guardians
- primary care physician
- pediatric ophthalmologist
- pediatric neurologist
- teacher of visually impaired
- occupational therapist
- physical therapist
- speech therapist
- teacher of the hearing impaired
- augmentative and alternate communication specialist
- orientation and mobility specialist
- feeding specialist

There is a wide range of challenges from cerebral palsy and CVI, each child is an individual. A plan includes identification of visual impairment, ophthalmologic examination documenting visual disability, questionnaires, observation of visual behavior and recommendations from educational professionals and other members on the patient's team should result in a treatment plan for each child with cerebral palsy and CVI.

Educational resources:
https://aapos.org/education/educational-resources/pediatric-low-vision-education