Subnormal vision

Detailed ophthalmic examination

Normal eye exam, disproportionately subnormal vision

Suspect CVI

Neurological evaluation

History

Neuroimaging

Abnormal anterior visual pathway

Diagnosis of CVI should be suspected in all children with unexplained bilaterally decreased vision
WHAT IS CEREBRAL VISUAL IMPAIRMENT OR CVI?

- Cerebral Visual Impairment (CVI) is deficiency in the function of vision

- Something wrong with visual pathways or processing centers in the brain (specifically those posterior to lateral geniculate body)

- Eye works fine but brain does not consistently understand or interpret what the eyes see
VISION ----- NEURAL ASSOCIATION

- DORSAL AND VENTRAL STREAM DYSFUNCTIONS

connects the occipital area with the posterior parietal cortex, which allows the mind to encompass the whole visual scene and to elect to pay attention to chosen components -- **WHERE**

connects the occipital and temporal lobe territories and subserves recognition of geometric and biological form, route finding, and visual memory -- **WHAT**

Brodsky MC. The apparently blind infant. In Pediatric neuro-ophthalmology 2016 (pp. 1-74). Springer, New York, NY
• Preterms- Myopia and its association with ROP
• Children with PVL who have escaped severe ROP- hypermetropia, often in combination with astigmatism
• Children with CP- Hypermetropia

OCULAR ASSOCIATIONS
Visual conceptual disorders
During the play you can also observe eye-hand coordination and awareness of directions, which are often problems for children with brain damage, even mild brain damage.

LEA Mailbox game:
The child is asked to drop a card through the slot of the LEA Mailbox Game. The child should demonstrate perception of the orientation of the slot by turning the card to the correct position before moving it half the distance to the slot (dorsal stream function).
Table 1.2  Neuro-ophthalmologic findings in cortical visual impairment versus subcortical injury (periventricular leukomalacia)

<table>
<thead>
<tr>
<th>Cortical</th>
<th>Subcortical</th>
</tr>
</thead>
<tbody>
<tr>
<td>Gaze deviation</td>
<td>Horizontal conjugate gaze deviation</td>
</tr>
<tr>
<td>Nystagmus</td>
<td>None or intermittent</td>
</tr>
<tr>
<td>Strabismus</td>
<td>Constant exotropia</td>
</tr>
<tr>
<td>Optic discs</td>
<td>Normal or mildly atrophic</td>
</tr>
<tr>
<td></td>
<td>Tonic downgaze</td>
</tr>
<tr>
<td></td>
<td>Latent or rarely congenital</td>
</tr>
<tr>
<td></td>
<td>Esotropia &gt; Exotropia</td>
</tr>
<tr>
<td></td>
<td>Hypoplastic or large cups</td>
</tr>
</tbody>
</table>

- Early PVL (<28 weeks)- Small discs, optic nerve hypoplasia
- >28 weeks- Normal discs with large cups- Pseudoglaucomatous discs
STRABISMUS IN CVI

Incidence:

• The rate of coexistent strabismus has been reported to be as high as 73% in patients with CVI

• Prevalence of strabismus in CP varies from 19% to 50%


STRABISMUS

Esotropia- most common type of strabismus (30-50%)

Exotropia- Constant, associated in more severely affected patients (10%)

Specific traits of Strabismus in CVI- *Angle variability* and *Shifting pattern*

Stable deviation on 3 follow up visits
Planned for surgery
Children with cerebral palsy and visual dysfunction caused by periventricular leukomalacia may have exo or esotropia, whereas those who have escaped cerebral palsy often present with esotropia.
Periventricular leukomalacia (PVL), an important neuroradiological sign of perinatal cerebral damage, causes visual impairment in children [1]. We retrospectively analysed the predisposing factors and ophthalmic manifestations in children<16y with MRI evidence of PVL in a tertiary care centre of Northern India. Records of 32 patients with PVL were analysed. Mean presenting age was 51.2 mo. Antenatal and perinatal history was positive in 43.7% and 96.8% children respectively Hypoglycemia, sepsis and birth asphyxia were cumulatively present in 20.6%. The major risk factor was perinatal hypoxia.
OUR DATA: 2011-2016

CVI with esotropia-23
Range of deviation- 12-60 PD
With myopia-3
With Nystagmus- 7
With Accomodative component- 2

CVI with exotropia-31
Range of deviation- 15-50 PD
IDS-10
V pattern-2
XT with DVD-2
Consecutive- 1
Strabismus and Ocular Motility.

- Motor evaluation revealed strabismus in 59 patients (67.04%).
- Exotropia was observed in 34 patients (57.62%) and esotropia was observed in 29 patients (49.15%).
- On further assessment, the deviation was variable in 7 patients (5 patients with esotropia and 2 patients with exotropia), intermittent in 6 patients (3 patients with esotropia and 3 patients with exotropia), and constant in the remaining 46 patients.

- The deviation was measured in 21 patients. The mean angle of deviation was 30.05 prism diopters for esotropia and 33.33 prism diopters for exotropia.
- An associated manifest disassociated vertical deviation was present in 8 patients and 2 patients had latent dissociated vertical deviation.
Parents came to us when he was 5 years of age with the following concerns:-

- Poor vision
- Squinting both eyes
- Maintaining head posture
- Very poor performance in school

BIRTH HISTORY
36 weeks born 2.5 kg
Did not cry at birth
NICU for 10 days
Perinatal Hypoxia, Infantile Arterial Ischemic stroke with seizures till 2 years of age

**MRI done** diagnosed HIE stage 2. Hyperintensity areas in bilateral cerebral hemispheres in fronto parietal and temporo occipital areas

Referred to ophthalmologist at 2 years as decrease vision and squinting noted by mother but fundus seen written within normal limits and no further treatment advised

At 3 years of age child fell down from stairs, suffered from right hemiplegia and facial palsy

Case of CVI
Vision 6/24 both eyes
Planning squint surgery at age 6 years
Why was surgery not done earlier?
Questions we ask ourselves?

THERE ARE NOT VERY CO-- OPERATIVE FOR MEASUREMENTS

DO CHILDREN WITH NEUROLOGICAL PROBLEMS HAVE DIFFERENT MOTOR OUTCOMES

DO THEY ACHIEVE BINOCULAR FUSION
N=70

Duration of follow up =10 years

**Management criteria:**

- Observe children with poor visual behaviour and with unstable deviation or shifting pattern
- Operate on them who have constant angle of >15 PD once they are able to follow faces or large toys irrespective of age (>4 years)

Repeated measurements- MUST- At least 4 stable and constant measurement before planning surgery

**CONCLUSIONS:**
16% showed spontaneous resolution
Not affected by cause of CVI
CASE

6Year/F
23/06/2010

• Having difficulty in maintaining concentration, eye-hand coordination, problem in focusing an object and inward deviation of RE

• 32 weeks prematurity, delayed developmental milestones and right sided hemiplegia

Refraction Atropine  
(OD) + 1.00DS  20/60  
(OS) + 1.00 DS  20/40

Parents consent taken
Orthoptics

- **Cover Test (D & N)**: Manifest LET to AET
- **Motility**: Full
- **PBCT (Distance)**: 45Δ BO
- **PBCT (Near)**: 45Δ BO
- **AHP**: Face turn towards right side

**Advised:**

Alternate occlusion 1:1

**Stable deviation on follow up visits**

**Planned for surgery**
S/P BE MR RECESSION 5 MM

With glasses

- **Vision (BE)** 20/30
- **Cover Test (D & N)** Small Residual Esotropia
- **PBCT(D & N)** 8Δ BO
- **4Δ BO test** Alt. suppression
- **Stereopsis** Grossly present with Lang Two Pencil test

05-08-2021
ROLE OF VISION REHABILITATION AND VISION THERAPY

THERAPIES POST SURGERY
<table>
<thead>
<tr>
<th>Name</th>
<th>MR No.</th>
<th>Diagnosis</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>P17/08/0260</td>
<td>Infantile Esotropia with CVI (BE)</td>
</tr>
</tbody>
</table>

**Pre-Op**

**Post-Op**
Long-term results of esotropia surgery in children with developmental delay

Zohar Habot-Wilner, MD,\textsuperscript{a} Abraham Spierer, MD,\textsuperscript{b} Irina S. Barequet, MD,\textsuperscript{b} and Tamara Wygnanski-Jaffe, MD\textsuperscript{b}

Effect: 2.26 degrees/mm

1. Less predictable surgical results
2. Overcorrections on long term follow up
3. Avoid surgery for smaller angles
4. Satisfactory results with additional procedures
**OUR STUDY: COMPARISON OF OUTCOMES OF INFANTILE ESOTROPIA WITH NEUROLOGICAL IMPAIRMENT COMPARED TO NORMAL CHILDREN**

<table>
<thead>
<tr>
<th></th>
<th>Neurological impairment (n=26)</th>
<th>Normal (n=102)</th>
<th>p value</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age of presentation</td>
<td>43.54±58.97</td>
<td>49.28±39.23</td>
<td>0.64</td>
</tr>
<tr>
<td>Age of surgery</td>
<td>47.58±58.54</td>
<td>57.26±39.51</td>
<td>0.39</td>
</tr>
<tr>
<td>Mean preop Spherical equivalent</td>
<td>1.23±2.36</td>
<td>1.83±1.93</td>
<td>0.002</td>
</tr>
<tr>
<td>Preop deviation</td>
<td>47.50±13.36</td>
<td>48.59±31.96</td>
<td>0.56</td>
</tr>
<tr>
<td>Followup period</td>
<td>23.31±19.66</td>
<td>28.33±26.32</td>
<td>0.29</td>
</tr>
</tbody>
</table>

![Chart showing percentage of neurological impairments]

- Cerebral palsy: 31%
- Birth asphyxia and HIE: 54%
- Seizures: 4%
In our study, we found surgical success rate of 61.23% in the neurological impairment group at a mean follow up of $23.31 \pm 19.66$ months.

In the group with no neurological impairment, surgical success was achieved in 70.6%. This was comparable between both the neurological impaired group and controls.

In the study done by Swaminathan et al, successful outcome was noted in 60% patients with developmental delay compared to 73.58% in control group.
### MOTOR SURGICAL OUTCOMES

<table>
<thead>
<tr>
<th>Motor surgical outcomes</th>
<th>Neurological</th>
<th>Non neurological</th>
</tr>
</thead>
<tbody>
<tr>
<td>Success(within±10 PD)</td>
<td>16(61.54%)</td>
<td>72(70.59%)</td>
</tr>
<tr>
<td>Consecutive Exotropia</td>
<td>2(7.69%)</td>
<td>9(8.82%)</td>
</tr>
<tr>
<td>Residual esotropia</td>
<td>8(30.76%)</td>
<td>21(20.59%)</td>
</tr>
<tr>
<td>Total</td>
<td>26(100%)</td>
<td>102(100%)</td>
</tr>
</tbody>
</table>

- Residual esotropia was treated with lateral rectus resection and consecutive exotropia was treated with medial rectus advancement.
- All achieved surgical success after second surgery.
RESULTS -- SENSORY

at a mean follow up of 23.31±19.66 months.

IOVS:
Repair of Strabismus and Binocular Fusion in Children with Cerebral Palsy: Gross Motor Function Classification Scale Fatema.F Ghasla et al

Restoration of binocular alignment and a degree of fusion is a realistic goal in the majority of strabismic CP children.
CONSECUTIVE XT ( OPERATED ET WHEN 2 YEARS OF AGE )
## SURGICAL DOSAGE

<table>
<thead>
<tr>
<th>AUTHOR</th>
<th>DOSAGE for BMR</th>
<th>MEAN FOLLOW UP</th>
<th>REMARKS</th>
</tr>
</thead>
<tbody>
<tr>
<td>Pickering et al</td>
<td>0.54 mm less (10.8%)</td>
<td>24 months</td>
<td>Increase in effect of the same amount of surgery averaged 5.28 prism diopters</td>
</tr>
<tr>
<td>JPOS 1994</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Wilner JH et al</td>
<td>0.84 mm less (16.8%)</td>
<td>17 months</td>
<td>Overcorrections on long term follow up</td>
</tr>
<tr>
<td>JAAPSO 2006</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Wilner JH et al</td>
<td>0.75 mm less (15%)</td>
<td>5 years</td>
<td>Effect: 2.26 degrees/mm</td>
</tr>
<tr>
<td>JAAPSO 2012</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>van Rijn LJ et al</td>
<td>Normal dose</td>
<td>2 years</td>
<td>Effect per mm of surgery in bimedial recession larger than generally experienced</td>
</tr>
<tr>
<td>Strabismus 2009</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Zehavi-Dorin T et al</td>
<td>0.66 mm less</td>
<td>4.6 years</td>
<td>Consecutive exotropia with time increased</td>
</tr>
<tr>
<td>Strabismus 2016</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>
Inward squint since age of 6 months
- Preterm history and history of delayed milestones
- BES MR recession done for 35 prism at age 2 (5.5mm)

Gradual outward deviation of right eye since one year

<table>
<thead>
<tr>
<th></th>
<th>OD</th>
<th>OS</th>
</tr>
</thead>
<tbody>
<tr>
<td>BCVA</td>
<td>6/18 (Allens)</td>
<td>6/18 (Allens)</td>
</tr>
<tr>
<td>Refraction</td>
<td>+2.5/- 4.0 @20</td>
<td>+2.0/-3.5 @170</td>
</tr>
</tbody>
</table>
Consecutive XT

Reported in 3% to 29% of all patients after surgical treatment of esotropia with higher incidences reported in studies with longer follow up.
PBCT – DVD 5 Prism OU

MR Advancement and LR recession done
Post op 4 weeks
A 6 year old girl child
Presented with decrease vision and
Squinting both eyes.

Mother says child falls down frequently
Not able to write properly
Attention disorder
Kept repeating words
Did not sit in one place for more than 10 minutes

MRI showed HIE

Vision 6/18 with exotropia both eyes.
1. On the Vineland Social Maturity Scale (VSMS), the child attained social quotient (SQ) of 74 indicating Borderline Level of Adaptive functioning.

<table>
<thead>
<tr>
<th>Total score</th>
<th>Chronological Age</th>
<th>Social Age</th>
<th>Social Quotient</th>
</tr>
</thead>
<tbody>
<tr>
<td>46</td>
<td>55 months (4.6 years)</td>
<td>40.4 months (3.4 years)</td>
<td>74</td>
</tr>
</tbody>
</table>

Pattern analysis (VSMS)

<table>
<thead>
<tr>
<th>Domain</th>
<th>Age(months)</th>
<th>SQ</th>
<th>Interpretation</th>
</tr>
</thead>
<tbody>
<tr>
<td>Self-Help General</td>
<td>36 months</td>
<td>65</td>
<td>Mild delays</td>
</tr>
<tr>
<td>Self-Help Eating</td>
<td>28 months</td>
<td>51</td>
<td>Moderate delays</td>
</tr>
<tr>
<td>Self-Help Dressing</td>
<td>32 months</td>
<td>58</td>
<td>Mild delays</td>
</tr>
<tr>
<td>Self-Direction</td>
<td>58 months</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Occupation</td>
<td>36 months</td>
<td>65</td>
<td>Mild Delays</td>
</tr>
<tr>
<td>Communication</td>
<td>40 months</td>
<td>73</td>
<td>Borderline</td>
</tr>
<tr>
<td>Locomotion</td>
<td>40 months</td>
<td>73</td>
<td>Borderline</td>
</tr>
<tr>
<td>Socialization</td>
<td>40 months</td>
<td>73</td>
<td>Borderline</td>
</tr>
<tr>
<td>Social Maturity</td>
<td>40.4 months</td>
<td>74</td>
<td>Borderline</td>
</tr>
</tbody>
</table>

*(Less than 25 Profound delay; 25-34 Severe Delay, 35-54 Moderate Delay, 55-79 Borderline delay; 80-89 Low Average; 90-109 Average; 110-119 High Average)*

### Before surgery

### After surgery
2. On Developmental Profile-3 (DP-3)

The child attained scores in below average range in all the domains of development. The child got a DQ of 74 indicating Borderline developmental Delay.

<table>
<thead>
<tr>
<th>Scale</th>
<th>Standard score</th>
<th>Age Equivalent</th>
<th>Descriptive category</th>
</tr>
</thead>
<tbody>
<tr>
<td>Physical</td>
<td>72</td>
<td>3 years 0 months</td>
<td>Borderline</td>
</tr>
<tr>
<td>Adaptive behaviour</td>
<td>70</td>
<td>2 years 10 months</td>
<td>Borderline</td>
</tr>
<tr>
<td>Social-Emotional</td>
<td>78</td>
<td>3 years 3 months</td>
<td>Borderline</td>
</tr>
<tr>
<td>Cognitive</td>
<td>76</td>
<td>3 years 6 months</td>
<td>Borderline</td>
</tr>
<tr>
<td>Communication</td>
<td>72</td>
<td>3 years 7 months</td>
<td>Borderline</td>
</tr>
<tr>
<td>GENERAL DEVELOPMENT</td>
<td>74</td>
<td></td>
<td>Borderline Development Delay</td>
</tr>
<tr>
<td>SCORE/DQ</td>
<td></td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

* (Greater than 130 Well above average; 116-130 Above average; 85-115 Average; 70-84 Below Average/Borderline; less than 70 delayed)
### On Child Behaviour Checklist (CBCL)

The child total behaviour problem score lies in Normal range. Her internalization as well as externalization behaviour scores are in Borderline clinical range.

#### The child got scores in clinical ranges in “Attention problems” scale.

<table>
<thead>
<tr>
<th>Syndrome scale</th>
<th>T-Score</th>
<th>Interpretation (Range)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Emotionally reactive</td>
<td>60</td>
<td>WNL</td>
</tr>
<tr>
<td>Anxious/ depressed</td>
<td>64</td>
<td>WNL</td>
</tr>
<tr>
<td>Somatic complaints</td>
<td>59</td>
<td>WNL</td>
</tr>
<tr>
<td>Withdrawn</td>
<td>69</td>
<td>WNL</td>
</tr>
<tr>
<td>Sleep problems</td>
<td>53</td>
<td>WNL</td>
</tr>
<tr>
<td>Attention Problems</td>
<td>70</td>
<td>Clinical</td>
</tr>
<tr>
<td>Aggressive behaviour</td>
<td>57</td>
<td>WNL</td>
</tr>
<tr>
<td>Internalizing</td>
<td>63</td>
<td>Borderline Clinical</td>
</tr>
<tr>
<td>Externalizing</td>
<td>61</td>
<td>Borderline Clinical</td>
</tr>
<tr>
<td>Total Problems</td>
<td>58</td>
<td>WNL</td>
</tr>
</tbody>
</table>

*50-64 Within Normal Limits (WNL); 65-69 Borderline clinical range; 70 and above clinical range

---

### Child Behaviour Checklist (CBCL)

The child total behaviour problem score lies in Clinical range. Her internalization behaviour score is also in clinical range. Her externalization behaviour score lies in Borderline clinical range.

#### The child got scores in clinical ranges in “Attention problems” and “Withdrawn” scales.

<table>
<thead>
<tr>
<th>Syndrome scale</th>
<th>T-Score</th>
<th>Interpretation (Range)</th>
</tr>
</thead>
<tbody>
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</tr>
<tr>
<td>Somatic complaints</td>
<td>53</td>
<td>WNL</td>
</tr>
<tr>
<td>Withdrawn</td>
<td>74</td>
<td>Clinical</td>
</tr>
<tr>
<td>Sleep problems</td>
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<td>WNL</td>
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<tr>
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<tr>
<td>Aggressive behaviour</td>
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<td>WNL</td>
</tr>
<tr>
<td>Internalizing</td>
<td>65</td>
<td>Borderline Clinical</td>
</tr>
<tr>
<td>Externalizing</td>
<td>61</td>
<td>Borderline Clinical</td>
</tr>
<tr>
<td>Total Problems</td>
<td>64</td>
<td>Clinical</td>
</tr>
</tbody>
</table>

*For syndrome scales: 50-64 Within Normal Limits (WNL); 65-69 Borderline clinical range; 70 and above clinical range*
Preterm CVI (HIE)
11 years
VA 6/12
No significant refractive error

CASE

PBCT – 30 XT with DVD 5 Prism OU

PBCT – 35 XT for D, 35 XT for N DVD 5 Prism OD, DVD 10 OS

PBCT – 45 XT for D DVD 5 Prism OU
OU LR recession
8mm with SR
recession 6mm
Boy 14 year old
Preterm 33 weeks –
Global developmental delay
Slow learner

Problems in visual perceptual skills and dyspraxia
VRC:
NEED TO WORK ON PERCEPTUAL SKILLS
MODIFICATION IN WRITING
START USING COMPUTER INSTEAD OF COPY TO WRITE
BRAIN GYM ACTIVITIES
Surgery done
RE LR RECESSION 6MM AND SR RECESSION 7MM, LE SR RECESSION 9MM)
Right eye IR plication
MIXED DEVIATIONS

- Upshoot in adduction
- “V” pattern (inferior oblique overaction)
- An isolated elevation deficit
- A monocular adduction, elevation, and depression deficit associated with partial ptosis

CONCLUSION

• CVI represents a spectrum of disability

• Diagnosis of CVI should be suspected in all children with unexplained bilaterally decreased vision, even when there are no overt neurological problems

• Significant benefit from specific intervention for ophthalmological abnormalities

• Ensure Multidisciplinary services

THANK YOU ......