Prematurity, the Eye and Vision

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The Systemic and Neurologic Impact of Prematurity

- Increased risk of
  - Chronic diseases eg asthma
  - Major neurological impairment
    - moderate/severe mental retardation
    - neurosensory disorders
    - epilepsy
    - cerebral palsy

Other Developmental Deficits

- Increased risk of
  - learning disabilities
  - ADHD
  - borderline mental retardation
  - behavioural problems, especially social and attention problems
  - specific neuropsychological deficits

Ophthalmic Manifestations of Preterm Birth

- Retinopathy of Prematurity
- Refractive Error
- Strabismus
- Cerebral Visual Impairment
- Colour Vision Defects
- Reduced Contrast Sensitivity
- Visual Field Defects
- Decreased Visual Acuity
• Myopia is well known to be associated with prematurity
  • “physiological” myopia
  • Myopia of Prematurity
  • myopia associated with severe ROP
  • Increased risk of all other refractive errors as well
  • overall risk of significant refractive error 29.6% in preterms, 6% in term
  • high hypermetropia
  • astigmatism and anisometropia


**Strabismus**

- Increased risk, but etiology complex
  - 14.7% incidence during infancy, but incidence remains high through 1st decade
- ET:XT ratio is 3:1 in FT, 1:1 in LBW kids
- Independent risk factors for squint in LBW kids include
  - Family history, ethnic origin, maternal age
  - Smoking, BW, general development quotient
  - Refractive error, anisometropia


**Cerebral Visual Impairment in Preterm Children**

- Brain injury in premmies due to mild-moderate hypoxia/hypo-perfusion
  - PVL
  - Periventricular haemorrhagic infarction
  - Germinal matrix haemorrhage
  - Cerebellar infarction

**Periventricular White Matter**

- Most common site of hypoxia/hypo-perfusion-related injury in prem babies
- Occurs in 32% of preemies
- Occurs between V24 and V34
- As well as optic tracts and radiations, cortico-spinal tracts run here
- Spastic diplegia occurs in 5-15% of preemies infants
- Most important in 50% of kids with spastic diplegia
- Neuro-imaging shows
  - Decreased density with irregular body and absence of lateral ventricles
  - Reduced volume of PVWM
  - Deep periventricular calcification or empty delta of ventricles
  - May also see delayed myelination and thinning of the corpus callosum

**Patterns of Functional Deficit in CVI**

- Impairment in CVI is variable, from NLP to normal VA but with cognitive visual dysfunction
- Cognitive visual dysfunction is a disorder of visual processing; may have normal VA and stereo
- Occurs with cortical lesions
- Visual function with cortex damage vs periventricular white matter damage
- Initial VA is similar (LP & occasional fixation in 22-38% & 49-50% respectively)
- 78% of children with damage to striate cortex improve but only 42% of those with PVL improve
- Significantly more strabismus, nystagmus & optic atrophy in PVL group
- VF defects common, especially in PVL
- Temporal lobe optic radiations less likely to be impaired in PVL
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CVI: associated ophthalmic and neurologic deficits

- ischemic injuries to retro-geniculate pathways causing CVI lead to different ophthalmic abnormalities if term onset v preterm onset
  - gaze disturbance
    - tonic downgaze in subcortical (preterm) visual loss
  - strabismus in ~80%
    - esotropia more common in subcortical (preterm) visual loss
    - exotropia more common in cortical (term) visual loss
  - nystagmus ~42%
    - now thought that nystagmus did not occur in retrogeniculate disease
    - now recognised that nystagmus occurs in a high frequency of children with PVL
  - optic nerves
    - normal in 54% of cortical visual loss but 29% of subcortical visual loss
    - optic nerve hypoplasia with pseudo-glaucomatous cupping in PVL
  - due to retrograde trans-synaptic degeneration of anterior visual pathways after serial insults have established a normal diameter
    - occurs only in immature visual system
    - results in nature retro-generative pathways do not lead to optic nerve cupping or damage

Retinopathy of Prematurity

- a vaso-proliferative disorder affecting low birth weight premature infants
- still a leading (preventable) cause of blindness throughout the world
- standardised international classification (ICROP) and robust evidence-based treatment guidelines (CRYO-ROP and ET-ROP) have led to improved outcomes in the developed world

Other Brain Injuries in Prematurity

- after profound hypotensive event or cardiopulmonary arrest
- deep gray matter and brainstem nuclei affected
- survival poor, but survivors often have
  - athetosis
  - quadripareisis
  - severe seizure disorders
  - mental retardation

Risk Factors for ROP: Historical Perspective

Cicatricial Changes in ROP

- Peripheral
  - vascular
  - vascular peripheral retinoschisis
  - abnormal non-dichotomous branching
  - circumferential connections between arcades
  - interarteriolar
  - retinal
  - pigmentary changes
  - disc retinoschisis
  - peripheral falciform
  - round mid-peripheral
  - lattice-like degeneration
  - radial breaks
  - TRD, RRD

- Posterior
What are the challenges that ROP poses?

- Examination / interpretation of findings require specialised training
- Infants need multiple examinations, requiring co-ordination between NICUs and ophthalmology screening programs
- Shortage of physicians available to treat ROP

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