CLINICAL PRACTICE GUIDELINE
Guideline coverage includes NICU KEMH, NICU PCH and NETS WA

Retinopathy of Prematurity (ROP) Screening, Treatment and Ophthalmology Consultations

This document should be read in conjunction with the Disclaimer

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Retinopathy of Prematurity (ROP) is a disorder of the eye, which results in the abnormal development of retinal blood vessels in the preterm neonate. The most significant risk factor is extreme prematurity. Those born < 31 weeks gestation or < 1250g are thought to be the most at risk.

The eye starts to develop at 16 weeks gestation, when the blood vessels of the retina begin to form at the optic nerve. The blood vessels gradually grow outwards towards the edge of the developing retina, supplying oxygen and nutrients. During the last 12 weeks of pregnancy the eye develops rapidly. When an infant is born full term, the retinal blood vessel growth is mostly complete. When an infant is born prematurely, the blood vessels have not reached the edges of the retina. Normal vessel growth may stop and the periphery of the retina may not get enough oxygen and nutrients.

It is believed that the periphery then sends out signals (by various mediators, such as Vascular Endothelial Growth Factor (VEGF)) to other areas of the retina for nourishment. As a result of this, new blood vessels begin to grow. These new blood vessels are fragile and weak and can bleed leading to retinal scarring. If the scarring shrinks, it can pull on the retina, causing it to detach. Retinal detachment is the main cause of visual impairment and blindness in premature neonates.

**Definition - International Classification of ROP (ICROP)**

There are five stages of ROP:

- **Stage 1**: Demarcation Line. This is a thin but definite structure that separates the avascular retina anteriorly from the vascularised retina posteriorly.
- **Stage II**: Ridge. It arises in the region of the demarcation line, has height and width, and extends above the plane of the retina.
- **Stage III**: Extraretinal Fibrovascular Proliferation (or neovascularisation) extends from the ridge into the vitreous.
- **Stage IV**: Partial Retinal Detachment.
  - **Stage IVA** – Partial retinal detachment that does not involve the fovea.
  - **Stage IVB** – Partial retinal detachment that involves the fovea.
- **Stage 5**: Total Retinal Detachment.

**Plus Disease**: Relates to the increased venous dilatation and arteriolar tortuosity of the posterior retinal vessels. Plus disease may also include iris vascular engorgement, poor pupillary dilation (rigid pupil) and vitreous haze.

**Pre-Plus Disease**: Relates to vascular abnormalities of the posterior pole that are insufficient for the diagnosis of plus disease, but in general, it is considered to be less than 2 quadrants of plus disease.

Most infants who develop ROP will have stage I or II, but a small number will worsen. Sometimes ROP can worsen very rapidly which can destroy vision.

Infants with ROP are also at increased risk of developing other eye problems later in life such as retinal detachment, myopia, strabismus, and amblyopia. In many cases these problems can be treated or controlled.
Risk Factors for Developing ROP

<table>
<thead>
<tr>
<th>Prematurity &lt; 31 weeks gestation</th>
<th>Resuscitation at birth</th>
</tr>
</thead>
<tbody>
<tr>
<td>Low birth weight &lt; 1250 grams</td>
<td>Multiple infections</td>
</tr>
<tr>
<td>Anaemia and Blood transfusions</td>
<td>RDS and prolonged ventilation</td>
</tr>
</tbody>
</table>

High levels of oxygen given to preterm neonates used to be an important risk factor but with newer technologies and monitoring of oxygen levels, this risk has diminished.

Screening Criteria

- All neonates born < 31 weeks gestation, regardless of weight.
- All neonates born < 1250 grams, regardless of gestation.
- Neonates will be screened at 4 weeks of age but **no earlier** than 31 weeks corrected gestational age.
- Neonates greater than 31 weeks gestation, born > 1250g, with additional medical concerns, will be screened at the discretion of the consultant and require a special consultation request.

<table>
<thead>
<tr>
<th>Gestation Age (GA) at Birth</th>
<th>Corrected GA (in weeks)</th>
<th>Age (in weeks)</th>
</tr>
</thead>
<tbody>
<tr>
<td>22 weeks</td>
<td>31</td>
<td>9</td>
</tr>
<tr>
<td>23 weeks</td>
<td>31</td>
<td>8</td>
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<td>24 weeks</td>
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<td>4</td>
</tr>
<tr>
<td>31 weeks</td>
<td>35</td>
<td>4</td>
</tr>
<tr>
<td>Older gestation &lt; 1250g</td>
<td>GA + 4 weeks</td>
<td>4</td>
</tr>
</tbody>
</table>

Adapted from American Academy of Paediatrics.
Transfer of Babies to Peripheral Hospital

- All neonates, which fit into the screening criteria, require a first review as an inpatient prior to transfer or discharge.
- If ROP is seen in the first review (or any subsequent reviews), this must be discussed and cleared with the treating consultant and ophthalmologist prior to transfer or discharge. A priority outpatient appointment (OPA) can be made at PCH as per the ophthalmologist request.
- If no ROP is seen and the neonate is suitable for transfer to a peripheral hospital, an OPA at PCH will be arranged for 38 weeks CGA.
- Low risk neonates (> 30 weeks) can have their imaging session bought forward to allow for transfer or discharged. A priority OPA will be made as per the ophthalmologist request.

Discharge Planning

- Neonates within the screening criteria should be flagged with the ROP screening team if transfer or discharge is approaching.
- Neonates within the screening program, that have not yet been ordered a 3 month review, are to be imaged the week of discharge.
- Neonates considered ‘safe’ by the screening ophthalmologist after the 38 week CGA review, can be ordered a 3-4 month OPA at PCH.
- If these ‘safe’ neonates remain as inpatients for > 4 weeks from the last date of imaging, the ophthalmologist is to be notified to see if repeat imaging is required prior to discharge.

RetCam Imaging

RetCam imaging is undertaken by trained RetCam nurses. Images are taken of the retina and sent directly to an ophthalmologist for review.

- Any neonate who fits the ROP screening criteria will be screened by RetCam.
- The procedure itself is short and with minimal discomfort to the neonate.
- Neonates who are deemed clinically unstable by the neonatologist are not imaged. The Ophthalmologist is to be informed and the neonate is to be screened at the earliest opportunity. If it is clear that the neonate will not be suitable for imaging for some time, the ophthalmologist will need to see the neonate in person for a BIO screen.

Dilacaine

- **Dilacaine Eye Drops** are used prior to examination to allow for clear examination of the fundus of the eye.
- Instil one drop of Dilacaine to each eye, 1 hour prior to examination and then again, 5 minutes after.
- Once Dilacaine has been administered, a biliband is placed over the baby’s eyes to protect from light sensitivity. This is to be kept in place for approximately 4 hours post examination, or longer if baby is showing signs of discomfort to light. During breastfeeds or cares with parents, the biliband can be removed, and then returned after these interactions.
Imaging Procedure

- The patent ID is checked and the neonate is identified on the RetCam software.
- Sucrose is administered and neonate securely swaddled.
- **Tetracaine Hydrochloride 0.5% Eye Drops** are distilled in each eye immediately prior to the procedure, to minimise discomfort.
- A speculum is used to allow a clear view of the retina with the camera lens.
- Right eye is imaged first.
- Polygel or Genteal Gel is administered for lubrication and to act as a bridging medium.
- The imaging session is videoed. Central, nasal, temporal, inferior and superior views are required.
- The speculum is removed, the gel is wiped away with sterile cotton balls and the RetCam lens is cleaned with 70% alcohol swab.
- The procedure is then repeated for the left eye.
- Once the procedure is complete, the neonate is returned to their cot/incubator and documentation completed.
- Still images are extracted from the video session.
- **If the images appear concerning, the ophthalmologist is to be contacted immediately to urgently review the images.**
- Images are transferred to an electronic reporting form and sent for review by the ophthalmologist. All images taken are reviewed by the ophthalmologist. The report is returned within 24 hours with follow-up instructions.
- The signed report is printed and filed in the patient’s notes.

**NOTE:**

- Any neonate not screened by the RetCam team, for any reason, is to have paperwork completed with documentation stating the reason why screening was not attended.
- The ophthalmologist is to be informed of any neonate not imaged and the reasons why.
- If an infection is present, a different speculum is used for each eye or a disposable speculum is used.
- Should the neonate’s clinical status deteriorate at any time, the procedure is immediately stopped. Continuing or rescheduling will depend on the neonate’s condition.
- Small red marks may be present on the neonate’s eye lids from the speculum, this usually resolves within 5-10 minutes.
- Parents are welcome to be present for the procedure.
Treatment Options

Laser Therapy
The most effective treatment for ROP is laser therapy; this burns away the periphery of the retina which is avascular. This reduces the production of VEGF, which is the driving factor for abnormal growth of blood vessels, which in turn reduces the risk of tractional retinal detachment. Laser therapy is indicated in Type 1 ROP, as defined by the ET-ROP trial (see reference). In brief, Type 1 ROP refers to any ROP with Plus disease, and Stage 3, Zone I without Plus. RetCam imaging can recommence one week post laser.

All babies requiring laser treatment will require ongoing follow up, and will require the ophthalmologist to clear them for discharge or transfer to peripheral hospitals.

Pre-Operative Care
- Follow the general pre-operative care guidelines. Pre-operative bloods are not a routine requirement for this procedure. An IV is usually inserted on the ward prior to theatre.
- Fast for 3 hours (for EBM/BF) or 4 hours (for formula).
- Instil one drop of Dilacaine into each eye every 30 minutes for two hours prior to time of surgery to dilate the pupil.

Post-Operative Care
- Routine Post-Operative Care. Connect to a cardio-respiratory monitor on return from theatre. Hourly observations - TPR and BP hourly for 4 hours, then 4 hourly for 24 hours.
- Check MR 842 operation summary for specific post-operative orders.
- Feeds can be recommenced once the infant is awake, following the anaesthetic.
- Commence eye drops approximately 4 hours post operatively. Chloramphenicol 0.5% and Flurometholone 1 mg/mL (FML) 1 drop to each eye 6 hourly, for 7 days as ordered.
- The Ophthalmologist will review this at the next ward round. Dilacaine dilation will be required for this.
- Continued Ophthalmology follow-up as an outpatient will be required on discharge home.

Avastin Treatment (Bevacizumab)
Avastin is a vascular endothelial growth factor inhibitor, which can be used to treat retinopathy of prematurity (ROP). In general, Avastin is used when the baby is too sick to have laser, or the disease is too posterior to allow laser to be done safely without risking damage to the macula or other significant visual structures. The BEAT-ROP trial suggested that the best indications for the use of Avastin is Zone I disease. There is also a tendency of late recurrence of ROP after Avastin. These babies need to be monitored for a longer duration after the treatment.

Avastin Procedure
- The Avastin must be ordered from pharmacy with at least 24 hours’ notice.
- The Ophthalmologist will bring all necessary equipment with him at the time of the procedure. It is done by the bedside with local anaesthetic, tetracaine.
• The baby will need to be swaddled and sucrose administered in preparation for the procedure. The infant needs to be held securely to minimise head movement and prevent any eye injury occurring during the procedure.
• The procedure is quick and conducted by the ophthalmologist only. It involves injecting the Avastin into the vitreous space under aseptic conditions.
• The ophthalmologist will then stipulate post procedure orders and when RetCam imaging can resume.
• All babies requiring Avastin treatment will require ongoing follow up OPA.

Ophthalmology Special Consultations
For any ophthalmic reviews not related to ROP, a special consultation form MR035 must be completed.
• The on call ophthalmology Registrar at PCH is to be phoned by the NICU medical staff, requesting a special consultation. Then the consultation form is then to be faxed to ophthalmology.
• If RetCam imaging is warranted – The ophthalmology Registrar will inform the NICU medical team, who will then inform the RetCam team.

References
3. Brian W. Fleck and Neil McIntosh. Retinopathy of Prematurity: Recent Developments
Related WNHS policies, procedures and guidelines

| Neonatal Medication Protocols: | Dilacaine Eye Drops  |
|                              | Tetracaine Hydrochloride 0.5% Eye Drops (Amethocaine) |
|                              | Chloramphenacol 0.5% |

| Neonatal Clinical Guidelines: | Pre-Operative Care |
|                             | Post-Operative Care |

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