Local Guideline and Procedure

John Hunter Children's Hospital
CHILDREN, YOUNG PEOPLE AND FAMILIES
NSW GOVERNMENT Health
Hunter New England Local Health District

Retinopathy of Prematurity (ROP) - Screening for and Management of in NICU

Sites where Local Guideline and Procedure applies	Neonatal Intensive care Unit, JHCH
This Local Guideline and Procedure applies to:	
1. Adults	No
2. Children up to 16 years	No
3. Neonates – less than 29 days	Yes
Target audience	All clinical staff who provide care to neonatal patients and non- clinical staff involved in outpatient eye appointments in NICU
Description	Provides information about diagnosis of ROP with the Retcam and management and treatment of ROP

Hyperlink to Procedure

Keywords	Cryotherapy, Hyperoxia, Preterm, Retina, Retinopathy of Prematurity (ROP), vascularisation
Document registration number	JHCH_NICU_18.01
Replaces existing document?	Yes
Registration number and dates of superseded documents	JHCH-NICU_18.01 June 2012

Related Legislation, Australian Standard, NSW Ministry of Health Policy Directive or Guideline, National Safety and Quality Health Service Standard (NSQHSS) and/or other, HNE Health Document, Professional Guideline, Code of Practice or Ethics:

- NSW Health Policy Directive 2014_036 Clinical Procedure Safety http://www0.health.nsw.gov.au/policies/pd/2014/pdf/PD2014_036.pdf
- NSW Health Policy PD 2005_406 Consent to Medical Treatment http://www.health.nsw.gov.au/policies/PD/2005/pdf/PD2005_406.pdf
- NSW Health Policy Directive PD 2007_036 Infection Control Policy http://www.health.nsw.gov.au/policies/pd/2007/pdf/PD2007_036.pdf
- NSW Health Policy Compliance Procedure Nurse and Midwife Initiated Medications PD2007_077:PCP4 Laser: Medical Laser procedures in GNAH facilities GNAH_0067.

Prerequisites (if required)	Ophthalmologist & Retcam operators with specialist training and education in Retcam use.
Local Guideline and Procedure note	This document reflects what is currently regarded as safe and appropriate practice. The guideline section does not replace the need for the application of clinical judgment in respect to each individual patient but the procedure/s **require mandatory compliance**. If staff believe that the procedure/s should not apply in a particular clinical situation they must seek advice from their unit manager/delegate and document the variance in the patients' health record.
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This document contains advice on therapeutics	Yes Approval gained from Local Quality Use of Medicines Committee on July 2015
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Note: Over time links in this document may cease working. Where this occurs please source the document in the PPG Directory at: http://ppg.hne.health.nsw.gov.au/

RISK STATEMENT

This local guideline has been developed to provide guidance to clinical staff in NICU to assist in assessment and management of ROP in the preterm infant. It ensures that the risks of harm to the baby's whilst caring for an infant being assessed and managed for ROP are identified and managed. *Any unplanned event resulting in, or with the potential for injury, damage or other loss to baby's/staff/family as a result of this management must be reported through the Incident Information Management System and managed in accordance with the Ministry of Health Policy Directive: Incident management PD2007_061. This would include unintended injury that results in disability, death or prolonged hospital stay.*
Risk Category: *Clinical Care & Patient Safety*

GLOSSARY

Acronym or Term	Definition
CGA	Corrected Gestational Age
EBM	Expressed breast Milk
PACS	Picture Archival Communication System
ROP/AP- ROP	Retinopathy of prematurity/Aggressive Posterior

GUIDELINE

This Guideline does not replace the need for the application of clinical judgment in respect to each individual patient.

OUTCOMES

1	Appropriate screening, treatment and follow up of at risk babies.
2	Early identification and management of ROP
3	Parents are fully informed of the progression of ROP and the screening/treatment process

Retinopathy of prematurity (ROP) and Retcam™ in NICU - One Page Summary and Checklist

(Ctrl+Click on **Coloured** words to jump to that section)

Introduction

top

Retinopathy of prematurity (ROP) is a disorder of the developing retina of low birthweight preterm infants (American Academy of Pediatrics (AAP), 2013), which involves disordered vascularization, cellular maturation and cellular differentiation. ROP is an important cause of visual impairment and can lead to blindness in low birthweight premature infants. The outcome for these infants can be improved if the disorder is detected by screening which allows appropriate treatment and follow-up (AAP, 2013; Pollan, 2009; Royal College of Ophthalmologists and British Association of Perinatal Medicine (RCO/BAPM), 1995 and Watts & Adams, 2000). This protocol for screening to detect ROP and its treatment is based on the Guidelines for Care around preterm birth (NHMRC 1996) and the Policy Statement by the AAP (2013).

Risk Factors

top

1. Preterm infants with a gestational age of 30 weeks or less (AAP, 2013).
2. Intrauterine growth restriction with a birth weight ≤ 1250g (Wright, Anderson, Walker & Lorch, 1998).
3. Preterm infants with a birthweight of >1250g or gestational age of >30 weeks with an unstable clinical course (AAP, 2013) such as:

    a. Hyperoxia – although the ROP epidemic in the mid - 20th century was due to excessive oxygen administration, this is rarely the sole cause in the modern era.
    b. Sepsis, blood transfusions, postnatal Dexamethasone use, germinal matrix intraventricular haemorrhage and Erythropoietin (in high cumulative doses), have been implicated as risk factors, but may reflect extreme prematurity and severity of illness rather than causation.

Consequences of Disease

top

In some infants this proliferative retinopathy progresses to inflammatory, haemorrhagic and ultimately fibrotic retinal scarring and detachment, which may lead to blindness (Watts & Adams, 2000). There are four stages of acute ROP. Adverse outcome is associated with stage 3 and 4 (see Appendix 1). The stages of ROP are outlined as:

Stage 1- Demarcation line – a flat simple border or line seen at the edge of vessels dividing vascular from avascular retina (a normal retina has a tenuous, non-linear, feathery border).

Image of early stage 1-2 showing demarcation line/ridge development

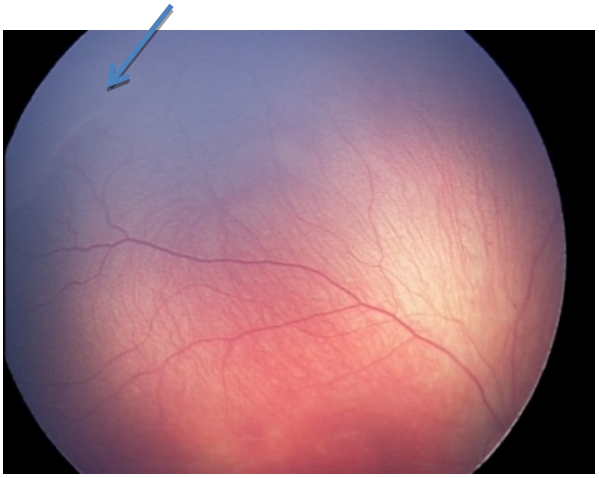
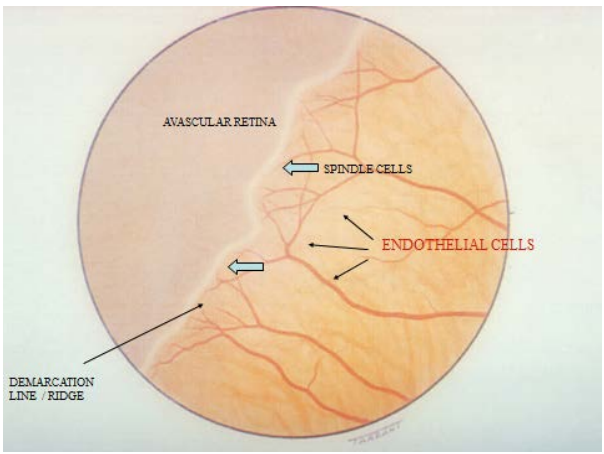


Diagram of demarcation line/ridge



Images taken by Retcam (Bruce McLeod, Sussex Eye Hospital. UK)

Stage 2- Elevated ridge – the line structure has acquired a volume and risen above the surface to become a ridge.

Stage 3- Neovascularization – a ridge is seen with extra-retinal fibrovascular proliferation. From the surface of the ridge, this extra-retinal tissue may extend into the vitreous.

Stage 3 can be further subdivided into mild, moderate or severe depending on the extent of new vessel growth projecting into the vitreous (Fraser Askin & Diehl-Jones, 2009).

Stage 4 - Subtotal retinal detachment caused by hardening or retraction of the scar tissue. This may be exudative and/or tractional, and could be partial or total detachment.

4A Partial detachment affecting the periphery of the retina. Macula not involved

4B Total detachment involving the Macula and Fovea, usually with a fold extending through Zones 1, 2 and 3 (Fraser Askin & Diehl-Jones, 2009).

Stage 5 - Total retinal detachment, with the retina assuming a closed or partially closed funnel from the optic nerve to the front of the eye.

Pre-'Plus' Disease- More arterial tortuosity and venous dilatation is seen than normal, but is not severe enough to be classified as 'Plus' disease.

Plus Disease

Plus disease is present when at any stage of ROP, the posterior veins are enlarged and arterioles are tortuous. The presence of plus disease increases the probability of rapid progression of the disease and significantly increases the chances of an unfavourable visual outcome (Fraser Askin & Diehl-Jones, 2009).

'Rush' Disease (aggressive posterior ROP, AP-ROP)

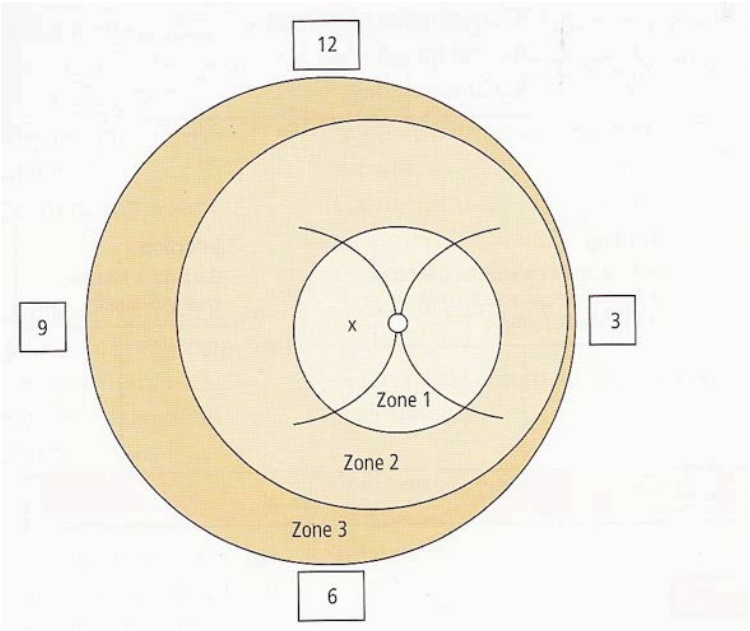
This is a rare and severe form of ROP with increased tortuosity and dilatation of vessels present in all 4 quadrants of zone 1 and sometimes zone 2. AP-ROP may not sequentially advance through stage 1 to 3, but often rapidly advance to stage 4 or 5.

Diagnosis

top

ROP is described according to the retinal zone involved (zones 1-3), the stage of disease (stages 1-5), the extent of disease (in clock hours) and whether there is 'plus' disease present. The aims of screening for ROP are to identify ROP which has the potential to reach stage 3 and that which may require treatment.

Zones - below is a schema of the retina of the right eye showing zone borders and clock hours which are used to describe the location and extent of ROP (AAP, 2013).



AAP 2013

Screening

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Screening for ROP should:

- be carried out on all infants who meet the criteria already described after day 28 of life and at least 31 weeks corrected gestational age (CGA)
- be repeated every 2 weeks or as required during the inpatient stay

Retcam images will be obtained by the Retcam operator for ROP screening and reviewed by an Ophthalmologist. All infants who are diagnosed with ROP will have regular reviews by an Ophthalmologist (AAP, 2013) and some Retcam images may be obtained for a general record of the disease progression. Even when no ROP is detected each infant who meets the criteria for ROP screening will have a physical indirect eye examination by an Ophthalmologist at least once during the inpatient stay (AAP, 2013). To ensure this is not missed all infants undergoing ROP screening will have indirect screening prior to transfer or discharge.

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Booking Eye checks:

top

- The Retcam™ nurses will maintain the eye appointments and book routine eye checks when they are due. However any other neonatal team member can add in patients for review to the eye book.
- On receipt of the review of the Retcam™ photographs the Retcam™ nurses will book any follow up appointments.
- When planning discharge, a referral will be made with a local Ophthalmologist for follow up if required.

Equipment required for nurse led ROP screening by Retcam™:

top

- Resuscitation equipment available and on standby.
- Eye drops- tetracaine 0.5%® cyclopentolate 0.5%® phenylephrine 2.5%®
- Lubricating eye gel -Polygel®
- Sucrose or Expressed Breast Milk (EBM)
- Retcam™
- Lid retractor
- Scleral depressor (only used by Ophthalmologist).

Lid retractor and scleral depressor MUST be sterilized and the sterilization identifier placed into the medical records after use.

Caution: Be aware of the contraindications for using the eye drops and ensure the baby is not on Beta Blockers (e.g Propranolol) prior to administration.

A standing order exists for the eye drops and must be signed by a medical officer within 24hrs. These eye drops should be checked by two Registered Nurses and documented in the patient's medication chart.

Procedure:

- Instil one drop of tetracaine 0.5%® to each eye. This is a local anaesthetic.
- Wait 2 minutes before instilling one drop of cyclopentolate 0.5%® and one drop of phenylephrine 2.5%® to each eye. Apply pressure to the lacrimal duct during and immediately after instillation to reduce systemic absorption (Laws et al., 1996). These are mydriatic and cycloplegic drugs, which will dilate the pupil and paralyse the ocular muscles of accommodation.

Prior to examination the infant will be wrapped and given oral pain relief in the form of oral Sucrose or a small amount of EBM. A second drop of tetracaine 0.5%® will be instilled into each eye. Ensure there are no bright lights on and insert the Lid retractors and Poly gel® (Lubricant) into the eye to be photographed. The Retcam™ will be used to take the photographs of each area of the infant's eyes. Following the procedure, the lid retractors will be removed and the patient observed for any complications or adverse events. The Retcam™ will be cleaned and the lid retractor sent for sterilization.

Note: Bright lighting should be avoided for two hours following the insertion of eye drops. The eye lids may be puffy for a while following the procedure and some infants may require additional monitoring for up to 2 hours post procedure.

The Retcam™ photographs will be uploaded to PACS by the Retcam™ nurses and reviewed by an Ophthalmologist. The Ophthalmologist may require the baby to have further photographs taken or an indirect eye examination. The Ophthalmologist will inform the Retcam™ nurses to organise this.

Documentation:

top

The Retcam™ nurse will document in the patient's medical records –

- that Retcam™ photographs have been taken
- how the procedure was tolerated by the baby

The Retcam nurse will also:

- update the eye book
- record the date of the next eye check
- add the Retcam™ results to the patient database

The Ophthalmologist will record in the infant's medical record her/his findings.

Treatment

top

The aim of treatment is to remove the stimulus for the vessel growth and to reduce the incidence of retinal detachment and blindness. Retinal ablation is effective treatment (AAP, 2013). Treatment should be undertaken as soon as possible, ideally within 2-3 days of the identification of threshold disease or the presence of plus disease in Zones 1 or 11 as follows:

- Zone 1 ROP: any stage with plus disease
- Zone 1 ROP stage 3 - no plus disease
- Zone 2 ROP: Stage 2 or 3 with plus disease (AAP, 2013).

A meta-analysis of the results of two randomised controlled trials shows that when ROP has reached threshold (stage 3+), treatment reduces the relative risk of unfavourable progression by 49% (RR 0.51, 95% CI 0.37 to 0.70) and of poor visual outcome in follow-up by 22% (CRPCG 1988, 1990, 1993; systematically reviewed by Watts 1992). Retinal ablation by laser (Laser is an acronym for Light Amplification by Stimulated Emission of Radiation) is now the therapy of choice as it is less invasive and less traumatic to the eye and produces less discomfort to the infant compared to retinal ablation using cryotherapy.

Involving the Parents

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It is important that all parents are fully informed about the need for ROP screening, the process of Retcam™ imaging, indirect ophthalmology examination and the results. It is important to emphasise that mild ROP is very common and spontaneously resolves without adverse sequelae in the vast majority of infants. If a parent refuses ROP screening after being fully informed about the screening process the matter should be referred to the Neonatologist for further counselling.

For parents of infant's with, or close to, severe ROP:

As soon as it is apparent that an infant has ROP, which is close to and/or likely to advance to stage 3, it is essential that the Ophthalmologist and the Neonatologist meets with the parents to discuss potential treatment, follow up and outcomes.

A fact sheet containing information about ROP sequelae, monitoring, treatment and outcomes will be given to the parents and any of their questions answered.

Follow-up Care

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Infants who have had ROP treatment require close follow up as determined by the Ophthalmologist. Parents of infants with severe visual impairment need support and advice about treatment options and referral to specific support groups for the visually impaired, such as State/Territory societies or institutes for the blind.

In addition, all infants with ROP of any stage require follow-up examination in the first four to six months of life and then at intervals determined by the findings. These babies are at risk of developing strabismus, amblyopia, high refractive errors and/or cataracts (AAP, 2013).

Flow Diagram for process for Retcam™

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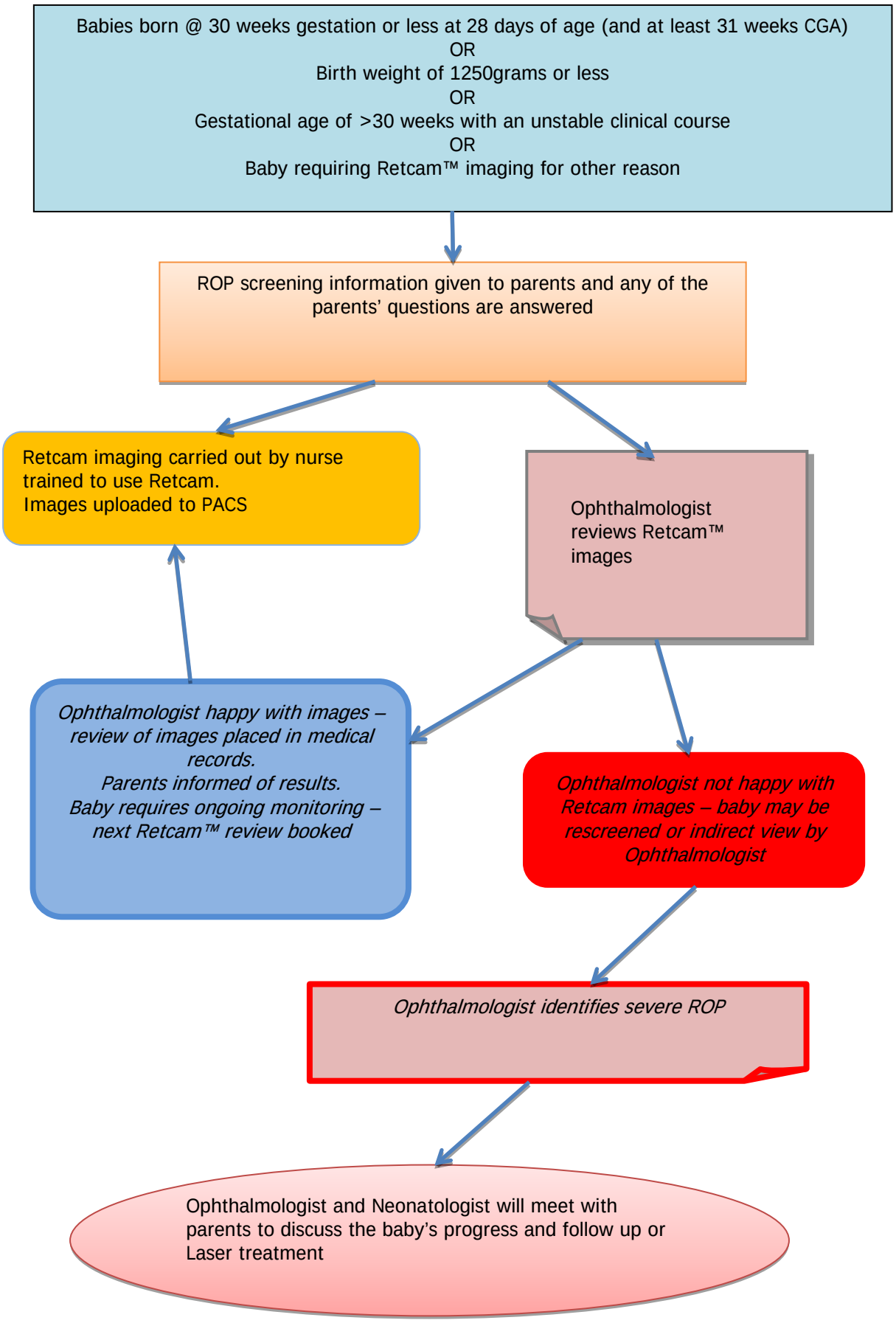
The following flow diagram outlines the process for Retcam™ imaging of the neonate in JHCH NICU. Note every baby who meets the criteria for ROP screening will have indirect Ophthalmology review at least once during the hospital stay.



Screening Frequency

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The reviewing ophthalmologist will advise the frequency of screening. The following recommendations of frequency are from the AAP (2013) and are used as a guide only.

Screening one week or less

Immature vascularization: Zone I no ROP
Immature retina extends into posterior Zone II, near the boundary of Zone I
Stage 1 or 2 ROP: Zone 1
Stage 3 ROP: Zone II
The presence or suspected presence of aggressive posterior ROP

Screening 1-2 weekly

Immature vascularization: posterior Zone II
Stage 2 ROP: Zone II
Unequivocally regressing ROP: Zone I

Screening 2 weekly

Stage 1 ROP: Zone II
Immature vascularization: Zone II- No ROP
Unequivocally regressing ROP : Zone II

Screening 2-3 weekly

Stage 1 or 2 ROP; Zone III
Regressing ROP: Zone III

References:

http://www.adhb.gov.nz/newborn/Guidelines.htm

American Academy of Pediatrics: Screening examination of premature infants for retinopathy of prematurity. Pediatrics 2013; 131(1):189–195.

Fraser Askin, D. & Diehl-Jones, W. (2009) Retinopathy of Prematurity Critical Care Nursing Clinics of North America 21:213-233.

International Classifications of ROP http://www.claritymsi.com <accessed 6/3/13>

Pollan, C. (2009) Retinopathy of Prematurity; An eye toward better outcomes. Neonatal Network 28:2. 93-101.

Royal College of Ophthalmologists and British Association of Perinatal Medicine (1995) Retinopathy of prematurity: guidelines for screening and treatment. The report of a joint working party. London: RCO/BAPM.

Retcam™™ http://www.claritymsi.com <accessed 6/3/13>

Retcam™™ (http://www.claritymsi.com/international/Retcam™_shuttle.html)

Watts, P. & Adams, G. (2000) In vitro fertilisation and stage 3 retinopathy of prematurity Eye 14, 330–333; doi:10.1038/eye.2000.82

Wright, K., Anderson, M.E., Walker, E. & Lorch,V. (1998) Should fewer premature infants be screened for Retinopathy of Prematurity in the managed care era? Pediatrics 102(1) 31-34.

RELATED LEGISLATION, DEPARTMENT OF HEALTH CIRCULARS, AREA POLICIES ETC:
Guidelines for Care around preterm birth (NHMRC 1996).

APPENDIX 1	International Classifications of ROP
APPENDIX 2	Fact Sheet: Eye Testing for Premature Infants
APPENDIX 3	Fact Sheet: Infants with Severe ROP
APPENDIX 4	ROP Screening Record

DEVELOPED BY: Shirley Graham NUM2 NICU JHCH

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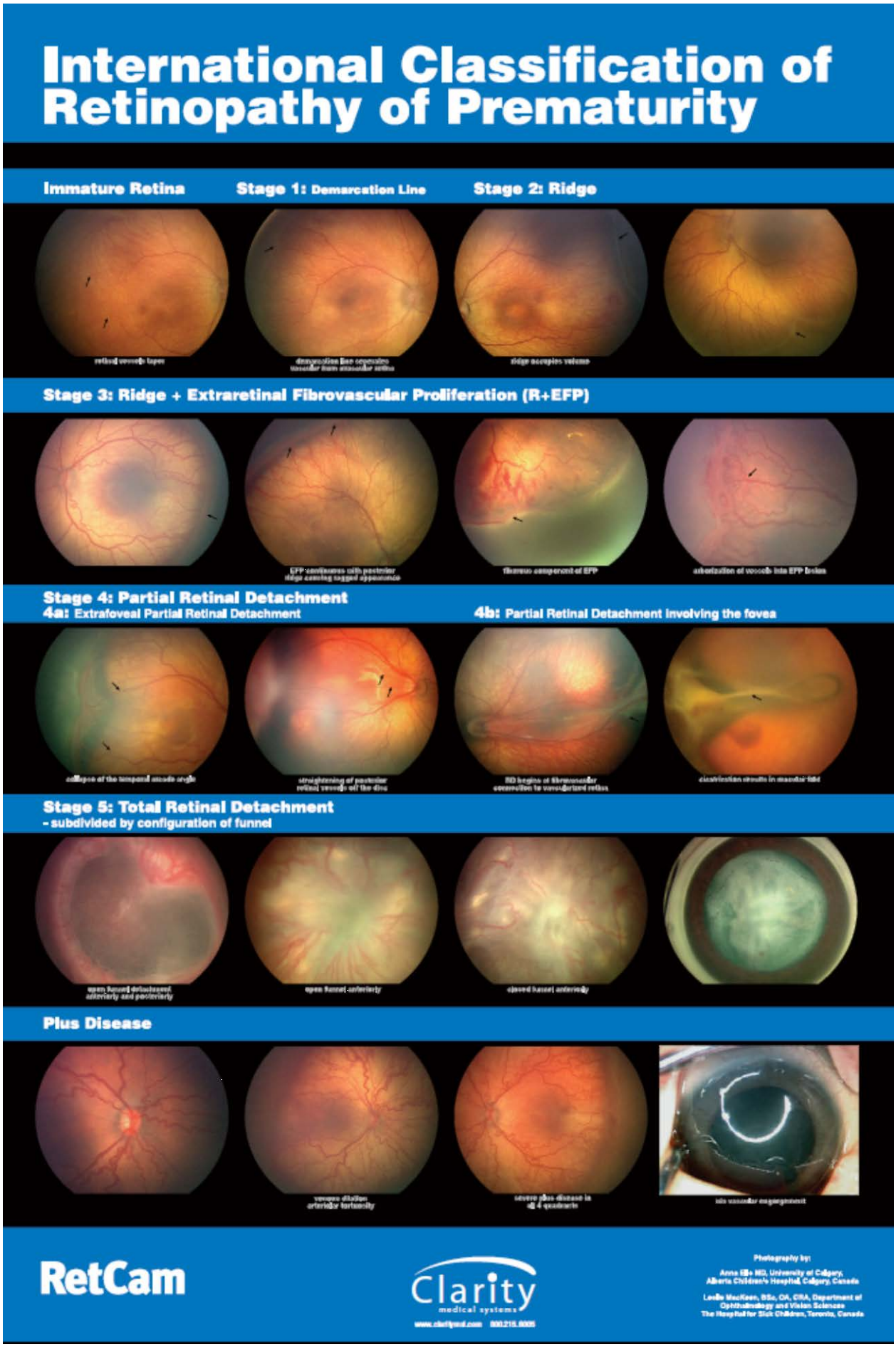
APPROVED BY: NICU Management Executive Committee 17/06/2015
Clinical Quality & Patient care Committee 24/11/2015

FEEDBACK

Any feedback on this document should be sent to the Contact Officer listed on the front page.

APPENDIX 1


International Classification of Retinopathy of Prematurity
Immature Retina
Stage 1: Demarcation Line
Stage 2: Ridge
Stage 3: Ridge + Extraretinal Fibrovascular Proliferation (R+EFP)
Stage 4: Partial Retinal Detachment
4a: Extrafoveal Partial Retinal Detachment
4b: Partial Retinal Detachment involving the fovea
Stage 5: Total Retinal Detachment
- subdivided by configuration of funnel
Plus Disease
RetCam
Clarity
medical systems

APPENDIX 2

FACT SHEET

Issued: October 2015

Eye testing for premature infants

The back part of the eye is called the Retina. It receives light and sends visual signals to the brain. The eye develops very quickly in the last 12 weeks of pregnancy. Therefore a baby born early has eyes that are still developing and is at risk of the blood vessels not growing normally. If the blood vessels do not grow normally they can detach the retina and cause severe visual problems. It is because of this that we monitor the developing retina in your baby's eyes.

Who needs eye checks?

All premature babies born at 30 weeks gestation or earlier and all babies weighing 1,250 grams or less will need eye checks. Also some other babies who were born after 30 weeks but before 32 weeks gestation will need to be screened if they have been unwell or your doctor is concerned.

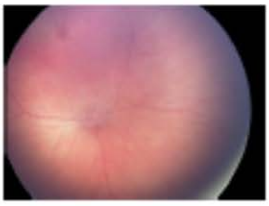
When will the eye check happen?

The first eye check will be done when your baby is 28 days old and at least 31 weeks corrected gestation. For example a baby born at 30 weeks will have an eye check at 34 weeks corrected gestation and a baby born at 25 weeks will have an eye check at 31-32 weeks corrected gestation.

What does the eye check involve?

The eye check involves eye drops being put into each eye at least 30 minutes before the eye check. These drops will numb the eye and dilate the pupil to allow us to take photographs of the back of the eye.

Once the pupils are dilated we use a machine similar to an ultrasound machine to take photographs of the eye. The following photograph is of a developing eye.



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APPENDIX 2 continued

Eye testing

An ophthalmologist (eye doctor) will look at the photographs, and if there are any concerns the ophthalmologist and/or the neonatologist (baby doctor) will let you know.

Does it hurt?

We give your baby some oral Sucrose and/or expressed breast milk for comfort as well as local anaesthetic eye drops to ease the discomfort. When the testing is complete a cuddle from parents helps too.

How often will it need to be done?

The eyes will need to be checked every two weeks until your baby's retina has reached full maturity (approximately when your baby was due to be born).

What if something is wrong?

The condition we are looking for is called Retinopathy of Prematurity (ROP) and it has several different stages. Mild ROP stage I or II generally resolves on its own, more advanced ROP may need treatment with Laser surgery. If your baby has ROP we will closely monitor this every week and if your baby needs treatment we will discuss Laser treatment with you. Although it is rare to have a baby with severe ROP we still monitor all babies so that we don't miss that one baby with problems. If your baby has any stage of ROP your baby will require follow up at approximately 4 months of age. This can be done in most towns across the region.

What if my baby needs Laser treatment?

If your baby has ROP that needs to be treated the eye doctor and the baby doctor will meet with you to tell you all about Laser treatment. The eye doctor does this treatment in theatre. Your baby will need to be cared for in the Intensive Care area of the nursery as babies may take some time to recover from both the surgery and the anaesthetic. If your baby needs Laser treatment it will be done within 2-3 days of recognising the need for surgery. This is to make sure we treat your baby before the ROP progresses.

How do I find out more information about Laser? You will be given another Fact sheet on Severe ROP, the baby doctor and eye doctor will have a chat with you and at any time you can also ask your baby's doctor.

Reference

American Academy of Pediatrics: Screening examination of premature infants for retinopathy of prematurity. Pediatrics 2013; 131(1):189–195

APPENDIX 3

FACT SHEET

Issued: October 2015

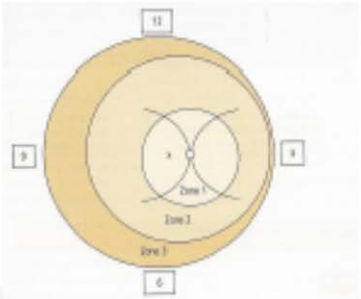
Information for parents of premature babies with severe Retinopathy of Prematurity

Severe Retinopathy of Prematurity (ROP) is often classified as stage III, IV or V. Severe ROP is rare but can cause major visual problems and sometimes blindness. The chance of these severe visual problems developing can be reduced by adequate screening and treatment.

What is severe ROP?

The abnormal blood vessel growth often starts as a bunched collection of blood vessels (a ridge) which becomes thicker and can cause a problem for future normal growth of vessels. Sometimes the ridge settles down and does not become severe ROP and sometimes it gets worse. Your baby will have eye checks every week and sometimes more often to monitor this.

The eye doctor describes the presence of ROP in Zones and stages. The stage refers to blood vessels growth, whereas the zone is the position of blood vessel growth on the retina. Severe ROP is when there is abnormal growth that is tortuous at the rim of the retina or any abnormal growth in the centre of the retina. If ROP is severe, treatment is needed within 2-3 days. The doctor will sit down and explain all of this for you. Please feel free to ask questions of your doctor at any time if you do not fully understand.



Why treat severe ROP?

If severe ROP is not treated the blood vessels will continue to grow abnormally and may burst and bleed. Treatment is to reduce this risk and stop the retina from becoming permanently damaged. If the retina becomes separated from the rest of the eye, eye sight will be lost and this cannot be repaired or replaced.

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APPENDIX 3 continued

Severe ROP

How do you treat it?

At this hospital we use Laser treatment to stop the abnormal growth of vessels. Laser is short for "Light Amplification by Stimulated Emission of Radiation" and Laser treatment is sometimes called 'retinal photocoagulation'. Laser is just a very intense light and it takes about 15-20 minutes for each eye. Your baby will have to go to theatre for Laser treatment, and this will require an anaesthetic. Your baby will be readmitted to intensive care after surgery for a recovery period.

Does it hurt?

No, your baby will be asleep when the laser treatment is being done. However your baby may have some discomfort on wakening and pain relief may be given then.

Your baby will receive some sucrose drops in the mouth as well as eye drops with local anaesthetic that will ease the discomfort

Is Laser treatment safe?

Laser treatment has been used for severe ROP in Australia since 1992 and in follow up of babies there have been no reports of any problems from the Laser treatment.

Before your baby has Laser treatment the baby doctor and the eye doctor will sit down with you to make sure you understand any complications of having an operation and what the side effects of Laser may be.

Will my baby have to go through this again?

Your baby's eyes will continue to be checked at regular intervals to make sure everything is going well. In a very small number of cases babies may need a second treatment with Laser. If your baby does require a repeat operation, it will be fully explained to you why this is required.

How do I find out more information about Laser?

Please ask your baby's doctor.

REMEMBER SEVERE ROP CAN BE TREATED

Reference

American Academy of Pediatrics: Screening examination of premature infants for retinopathy of prematurity. Pediatrics 2013; 131(1):189–195

APPENDIX 4

ROP screening record

Name: Attach patient ID sticker

MRN:

Gestational Age at birth:

Birthweight:

Parent/Carer contact number:

Relevant history:

Date	CGA	Retina Mature	ROP screening results	Comments	Next review

Follow up with _____

Date: Time: