THE ROYAL COLLEGE OF OPHTHALMOLOGISTS
BRITISH ASSOCIATION OF PERINATAL MEDICINE

RETINOPATHY OF PREMATURITY:
GUIDELINES FOR SCREENING AND TREATMENT

The Report of a Joint Working Party

1995
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1. **SUMMARY**

Following the report that severe retinopathy of prematurity (ROP) can be treated effectively a working party of the Royal College of Ophthalmologists and the British Association of Perinatal Medicine was convened in 1990 to draw up guidelines for screening\(^1\). These have been revised by a reconvened working party and the document expanded to include other relevant issues such as the practicalities of treatment, counselling and end-stage retinopathy.

- Screening is recommended for all babies at risk of severe ROP, i.e. those of birthweight \(\leq 1500\text{g}\) and/ or \(\leq 31\) weeks gestational age.

The aim of screening is to identify severe ROP (stage 3) which may require treatment, or, in a baby due to be discharged to home or to another hospital, any ROP which has the potential to become severe.

- The first examination should be between 6 and 7 weeks postnatal age, and subsequent examinations continued until vascularisation has progressed into zone 3 when the risk of stage 3 ROP has passed.

- As the window of time available for treatment is very short, examinations should be undertaken every two weeks.

- Treatment is currently undertaken when threshold stage has been reached.

Threshold ROP is defined as:

Stage 3 ROP:
- involving 5 or more contiguous, or 8 or more cumulative, clock hours
- in the presence of congestion of the posterior pole vessels – ‘plus’ disease.

- Treatment can be by cryotherapy or laser in the neonatal unit.

- Parents have a right to know what may befall their baby, and it is important to provide balanced information. This should be given to parents of babies:
  - at risk of developing any ROP
  - who are close to or have severe disease or requiring treatment
  - blinded by ROP

- Despite meticulous clinical care babies are occasionally blinded by ROP. That is not the end of the road and there is still work for the clinician to do:
  - counsel on the role of vitrectomy
  - manage the disorganised anterior segment
  - initiate and actively participate in the care of the visually-impaired child.

The screening and management should be undertaken or supervised by senior ophthalmologists with a specific interest in this condition. It is recommended that one or two consultants in each area should gain expertise and supervise training.
Referencing here is not exhaustive, with preference being given to recent reviews. With certain exceptions citations are at the commencement of each section rather than at specific issues.

2. BACKGROUND
Survival rates for preterm babies have increased significantly over the last 40 years, from about 5% to 65% for babies with birthweight less than 1000g and from about 35% to 90% between 1000g and 1500g birthweight. Consequent upon this improved survival, the number of babies with retinopathy of prematurity (ROP) has risen. It is not our purpose here to enter a detailed discussion of its pathogenesis but to recognise that despite meticulous neonatal care severe disease still occurs and at present ROP is not entirely preventable.

In the past the ophthalmologist could neither prevent nor effectively treat ROP and therefore the medical responsibility for this condition rested with issues related to causation. The results of the US-based Multicenter Trial of Cryotherapy for ROP2-5 have changed this position by demonstrating that the unfavourable outcome of ‘threshold’ stage 3 disease can be significantly reduced by active treatment. Therefore the early identification of ROP by screening should be standard practice. Although the number of infants blinded by ROP is fortunately relatively small, this represents a great number of years of disability, which in many cases is unnecessary.

ROP screening and treatment have been the subject of several recent publications 6-17.

2.1 Incidence and natural history of ROP
Both the incidence and severity of acute ROP rise with decreasing gestational age and whilst estimates vary13,15,18-26, about 30% to 60% of babies weighing less than 1500g develop some degree of retinopathy. Severe disease (stage 3 or above) is virtually confined to those infants of birthweight <1500g and <31 weeks gestational age (GA)*

Screening protocol design is facilitated by the finding that the onset and progression of ROP are both related more to the stage of development of the infant than to factors such as postnatal age, severity and timing of adverse neonatal events and oxygen therapy13,15,27-29. Thus the smaller, more immature, baby will develop retinopathy at a later postnatal age than his/her larger counterpart, irrespective of the general condition.

Most acute ROP develops between about 32 and 44 weeks postmenstrual age (PMA). ROP is rare before 31 weeks, and stage 3 occurs between about 34 and 42 weeks PMA. Disease commencing after 36 weeks PMA is unlikely to become severe.

The location and extent of the lesion indicates its potential to become severe. Thus the more posterior the location of the retinopathy and the greater its extent the faster its progression and the greater the propensity of the lesion to become severe15,28,30.

*Gestational age is the period in utero and is calculated from the first day of the last menstrual period. Once birth has occurred gestational age is inappropriate, and postnatal, postmenstrual age, and postconceptional age are used. Postmenstrual age is used to denote gestational age plus postnatal age, in weeks. Postconceptional and postmenstrual age are used synonymously.
Stage 3 acute ROP may, or may not, resolve. The term threshold denotes the ROP stage at which spontaneous and complete resolution is unlikely and the risk of blindness is predicted to be close to 50\%.

Threshold is defined as \(^{31}\):
Stage 3 ROP:
- involving 5 or more contiguous, or 8 or more cumulative, clock hours
- in the presence of congestion of the posterior pole vessels – ‘plus’ disease.

‘Plus’ disease is an indicator of activity. In order of severity ‘plus’ signs include:
- engorgement and tortuosity of the posterior pole retinal vessels
- iris vessel engorgement
- pupil rigidity
- vitreous haze

By the time vitreous haze and sometimes the iris changes have been reached the eye is untreatable.

Where the maximum stage reached is 1 or 2, all cases of acute ROP undergo spontaneous resolution and do not result in visually disabling sequelae \(^{32,33}\). Screening for amblyopia falls outside the remit of these guidelines.

3. SCREENING FOR ROP

3.1 Aims of screening
To identify:
- ROP which has the potential to reach stage 3.
- Severe (stage 3) ROP which may require treatment.

3.2 The population at risk
About 6,600 infants of birthweight <1500g are born in the UK each year with a survival rate of 80\% (5,280) and an incidence of stage 3 ROP of approximately 8\% to 10\% (about 450 infants). The risk of severe ROP in larger or more mature babies is extremely low and for these screening cannot be justified.

3.3 The ophthalmic examination
Three aspects are important:

A. Timing of the examination
B. Determining the severity
C. Identifying the location (by zone and extent by clock hours)

A. Timing of the examination

Principle: The onset and progression of ROP are both determined predominantly by PMA rather than by neonatal events and this can be used to predetermine the time of
screening examinations. The word *predominantly* must be emphasised, for severe ROP has a tendency to progress more rapidly than mild disease \(^{13,15,27-29}\). In addition,

- ROP onset is rare before 30 weeks PMA. A normal examination undertaken very early may have no value.

- The mean age for the onset of threshold ROP is around 37 weeks PMA, but can occur earlier in rapidly progressive disease, especially in zone 1 in the presence of ‘plus’ signs \(^{25}\)

- The time available for treatment is very short (around 2 weeks) and screening examinations must be timed accordingly \(^{2,25,30,34}\)

- The timescale for developing serious ROP may necessitate occasionally examining the very sick neonate. Babies in incubators, or receiving supplemental oxygen, must still be examined

- *A first examination should be undertaken in hospital.* Visits to outpatient department greatly inconvenience the family, and failure to attend at the appropriate time, for whatever reason, may have disastrous consequences

- For screening purposes it is not necessary to review all babies with stages 1 and 2 until the point of complete resolution, and it is adequate to ensure that this process is well underway

**B. Determining the severity**

The International Classification for Retinopathy of Prematurity (ICROP) \(^{31,35}\) to record clinical findings (Appendix 1) should be used.

- Signs of ‘plus’ disease, especially in the anterior segment, are all ominous signs and the pupil which fails to dilate may well be the one harbouring serious ROP

- Persistence of the tunica vasculosa lentis after its usual time of regression by 34 weeks PMA may be an indicator of severe disease.

**C. Identifying the location (by zone and extent by clock hours)**

**Principles:**

- The more posterior the retinopathy the greater the propensity for progression to severe ROP \(^{30}\)

- Site of onset and extent of involvement can be useful indicators of subsequent progress \(^{15,28,30}\)
In addition:

- Zone 1 very frequently does, and zone 2 ROP may, progress to stage 3
- The greater the number of clock hours the greater the likelihood of progression
- Any ROP in zones 1 and 2 should be kept under observation until resolution is well underway
- ROP confined to zone 3 alone rarely progresses to stage 3
- ROP in the most immature neonate often starts in the nasal retina
- For those without ROP, once the vessels have entered zone 3, the risk of serious stage 3 has passed and screening may cease
- Retinopathy involving the nasal retina (i.e. zone 2 disease), and/or the vertical regions all have a greater propensity to progress to stage 3 than ROP confined to the temporal retina alone
- The examiner can only be certain that ROP is located in zone 3 if the nasal retina is fully vascularised (see examination technique later)

3.4 Which babies should be screened?
- ≤31 weeks gestational age
- ≤1500g

3.5 Examination protocol
A) Commencement
All babies must be screened at 6-7 weeks postnatal age. For the more mature baby one examination before discharge may be all that is necessary even if undertaken before 6 weeks because this gives enough information about retinal vascular development to indicate the need for further examinations.

B) Subsequent examinations
At least every two weeks until vascularisation has progressed into zone 3.
- Because of the short time available for treatment, if stage 3 is imminent or present it is sometimes necessary to examine a baby more frequently.

C) Babies due for transfer or discharge to home
To ensure that the screening process is completed, the appropriate arrangements must be made for babies due to be transferred to another hospital or discharged home. Neonatal unit staff must inform the receiving hospital that the screening process requires to be completed, or, if the infant is discharged home, the appropriate follow-up appointment must be made.
D) **Follow-up**
As dictated by clinical criteria on an individual basis.

The long-term review of preterm infants for amblyopia and other visual pathway defects and strabismus is beyond the scope of these guidelines, although the incidence of the latter is higher than in the full-term population\textsuperscript{32,33}. Review of all stage 3 infants is advisable, at least in the preschool years, as the incidence of strabismus and other problems is particularly high in this group.

3.6 **Examination technique**
The following are recommend:

A) **Pupil dilatation by either**

- i) Guttae cyclopentolate 0.5%
- or
- ii) Guttae cyclopentolate 0.5% with guttae phenylephrine 2.5%. As the dilatation may last more than 12 hours, weaker solutions may be used, such as cyclopentolate 0.2% in combination with phenylephrine 1.0%.

All mydriatic eyedrops should be instilled at least 30 minutes prior to examination.

B) **Indirect ophthalmoscopy with a 28 or 30 dioptre lens**
For routine screening disease, the use of an eyelid speculum and scleral indentor is no mandatory. Nevertheless, they are widely used, and permit the scrutiny of the most peripheral regions of the retina, which is important in determining whether the nasal retina is fully vascularised (i.e. zone 2 or 3). Preferably, topical anaesthetic eyedrops are instilled prior to inserting the eyelid speculum.

3.7 **Record keeping**
Using ICROP\textsuperscript{31,35} record the following (Appendix 1):

- ROP by:
  - severity by stage
  - location by zone
  - extent in clock hours
  - ‘plus’ disease

- Other clinical signs
  Record also any changes in the cornea, anterior chamber, iris, pupil, lens and vitreous.

- Signs of regressed ROP

- Arrangements for review should be indicated in the hospital records.
3.8 Parental information
It is helpful to explain to parents the reason for ophthalmic screening. Also provide information, preferably separately, for those whose babies are likely to, or already have, developed severe ROP (see Section 5 on counselling and Appendices 2 and 3).

4. TREATMENT OF THE ACUTE ROP
The Multicenter Trial of Cryotherapy for ROP has demonstrated that cryotherapy significantly improves the favourable outcome of severe ROP. More recently, portable indirect ophthalmoscopic argon or diode lasers have also been used with similar success. The outcomes of these treatments have not been reviewed here as the purpose of these guidelines is to focus on the practical aspects of treatment deliver.

4.1 Rationale for treatment
ROP is a disorder of the immature retinal vasculature. The stimulus for abnormal vessel growth comes from the peripheral, yet to be vascularised, retina. The fundamental principle of treatment is to remove the stimulus for vessel growth, i.e. ablate the peripheral avascular retina.

4.2 Criteria for treatment
The current indication for treatment is threshold ROP. These guidelines reflect prevailing safe practice and therefore cannot, at this stage, deal with treatment of ROP at earlier stages although future criteria for intervention may change. This applies particularly to zone 1 disease as many ophthalmologists now treat any stage 3 in this location.

4.3 Timing
Timing is critical because once the vitreous has become involved, or cicatrisation has commenced, retinal ablation by either cryotherapy or laser is ineffective. Treatment should therefore be undertaken as soon as possible, ideally within 2-3 days of the identification of threshold disease.

4.4 Preparing the parents
This is a difficult time for parents, who may be recovering from the stress of their baby’s acute illness. The news that their baby has a potentially blinding condition which requires urgent treatment must be handled with the utmost consideration. This is helped by the provision of written information explaining screening and severe ROP – see Appendices 2 and 3.

4.5 Preparing the baby
Close supervision and monitoring throughout the procedure can best be provided in the neonatal unit. Cryotherapy and laser treatment are painful and/or lengthy procedures and good analgesia and sedation are essential. A venous cannula for drugs and facilities for artificial ventilation are also essential. The optimal condition is for the baby to be intubated and given artificial ventilation. Experienced staff must be in attendance throughout and cardiorespiratory monitoring continued until the baby has fully recovered.
4.6 Preparing the eye
Dilate the pupils fully, bearing in mind the potential systemic toxicity of these topical agents. Do not use subconjunctival or retrobulbar anaesthetics\textsuperscript{46}.

4.7 Treatment overview
Both cryotherapy and laser are effective in the treatment of severe ROP. In this rapidly advancing field, the relative advantages of each modality have yet to be determined, in either the short- or the long-term.

- Cryotherapy applications are applied contiguously\textsuperscript{2,17,34} whereas laser applications are spaced one half burn-width apart\textsuperscript{41,42}.

- Cryotherapy requires ocular manipulation, is painful, and systemic upset is more likely than with laser, but treatment time is less.

- Cryotherapy can be used where the retina cannot be visualised, when the pupil will not dilate, or haemorrhage obscures the view.

- Laser is preferable when the infant is unstable, can be used simply when retreatment is necessary, and is more portable than cryotherapy equipment, thus facilitating the treatment of infants who cannot be transferred to a major centre.

- Laser wavelength must also be considered. Diode laser can be used through cloudy media. Diode laser is less likely to be absorbed by the ocular tissue and hence less prone to cause anterior segment damage and is probably the preferred mode.

The long-term sequelae of either cryotherapy and laser on ocular growth, and the risk of late retinal detachment, are not yet fully understood.

4.8 Cryotherapy
- Cryotherapy is applied trans-sclerally to the avascular zone anterior to the acute ROP lesion. Use a retinal probe or one specially designed for the purpose (e.g. Schulenburg probe), and ensure that the probe shaft is guarded to protect the eyelids.

- It is recommended that the entire $360^\circ$ of the circumference of the globe is treated under direct visualisation. As a guide, this usually means two rows temporally and one nasally. Applications limited to the areas of stage 3 have been reported\textsuperscript{47}.

- Start treatment in the temporal retina first where access is better

- Avoid applications to the ridge itself. The conjunctive may need to be opened by a radial incision if the probe does not reach far enough back

- Allow defrosting before removing the probe to avoid fracturing the choroid

- Between cryotherapy probe applications rest from time to time to prevent central retinal artery occlusion
4.8.1. Complications of cryotherapy
Systemic complications described include bradycardia, cyanosis and respiratory depression\textsuperscript{17,46}. Ocular complications include eyelid oedema, lacerations and haemorrhage of the conjunctive, preretinal and vitreous haemorrhage\textsuperscript{3,14,17}.

4.9 Laser therapy
Both argon-green and diode-red wavelengths can be delivered through a portable indirect ophthalmoscope. Instead of confluence, leave a small space of half a retinal burn between lesions\textsuperscript{41}.

- Laser is simpler to deliver to posterior disease, which carries the worst prognosis.

4.9.1. Complications of laser therapy
Laser complications include corneal, iris and lens burn, and cataract has been reported. Argon may be absorbed by the tunica vasculosa lentis. Retinal or vitreous haemorrhage may occur.

4.10 Postoperative management
Mydriatic (not atropine), antibiotic and sometimes steroid eyedrops are instilled for a few days postoperatively.

- Cryotherapy reactions appear within a few days
- Examine the retina in about 5-7 days, by which time ‘plus’ disease should show signs of subsiding

With effective treatment, retreatment is not often needed. However, if it is, it should be undertaken as soon as it is obvious that regression is patchy and that there is still active retinopathy with the potential for traction. Temporal disease is of more concern than that nasal to the optic disc. In the absence of active ROP it is probably not necessary to treat all skip areas. Resolution may take a few days longer following laser, but the need for careful monitoring with regard to possible retreatment is identical.

4.11 Long-term follow-up
All infants with stage 3, and those who have been treated, should be kept under review for at least the preschool years to monitor the development of vision, refractive status and ocular motility.

5. COUNSELLING
The parents of a very preterm infant have much to cope with. They have a right to know what may befall their baby. Providing sensitive, balanced information is an essential component of this process to enable parents to be involved in decision-making\textsuperscript{48}. Information is required at several stages. The purpose of this statement is to highlight this issue, and not to provide full details which will dictated by local circumstances (see Appendices 2 and 3).
Sample written information is given in Appendices 2 and 3. The purpose of keeping documentation on screening and severe disease separate is not to withhold information but to avoid causing unnecessary anxiety for the vast majority of parents whose babies will never develop visually threatening disease.

5.1 **For parents of all babies at risk**
Written information should be provided as part of the general information provided by the neonatal unit for parents of premature infants. This might state that mild ROP is very common and spontaneously resolves without adverse sequelae in the vast majority.

5.2 **For parents of infants with, or close to, severe ROP**
As soon as it is apparent that an infant has ROP which is close to and likely to advance to stage 3, it is preferable that the ophthalmologist personally discusses the issues with the parents. It is recommended that a member of the neonatal unit staff who knows the family, such as the consultant paediatrician or the senior neonatal nurse, is present so that post-interview queries can be dealt with. Remember that severe ROP often occurs just when parents are beginning to relax for the first time after stressful weeks of uncertainty.

Be open and keep parents fully and frequently informed. The complexity of accurately monitoring visual functions and the difficulty of predicting the future must not be underestimated. Nevertheless, measuring visual functions at each stage of the review process does enable the clinician to advise parents on progress.

5.3 **For parents of infants with advanced cicatricial ROP**
Despite the tragedy the need to keep in close contact with the family cannot be over-emphasised. Generally, parents can accept with time that not all disorders can be successfully treated, but they cannot accept lack of interest or care (see below for details of treatment of end-stage disease).

*The need for a balanced and sensitive approach at all stage is emphasised.*

6. **MANAGEMENT OF THE ADVANCED DISEASE**
Neither cryotherapy nor laser are a panacea, and despite meticulous management, for a number there are sequelae, some of which are serious. Naturally the parents will want to know if anything can be done. There are several aspects to consider.

6.1 **Vitreoretinal surgery**
This is a most controversial topic. Anatomical retinal alignment can be obtained in a significant proportion of cases, but to date the visual results are extremely dismal with almost all infants gaining no pattern recognition. However, a dilemma is created because a few surgeons in the USA have reported slightly more favourable results. Parents may well ‘clutch at straws’ to obtain any vision for their baby. The clinician counselling in this situation should point out the distinction between anatomical and functional success and advise the parents that vitrectomy is major ocular surgery, and, if the eye has any function, this is place at risk by such a procedure. In the UK currently, because of the poor functional results, vitrectomy is not recommended.
6.2 Disorganised anterior segment
Due to anterior iris-lens displacement, children with advanced cicatricial ROP often have shallow or flat anterior chambers\textsuperscript{53}. Glaucoma may develop. Lens extraction, prophylactic or reactive, should be considered. Band keratopathy may require treatment.

In the presence of an obliterated anterior chamber where glaucoma is likely, prophylactic lensectomy should be considered.

6.3 Care of the visually-impaired child
This cannot be considered in detail here, but the ophthalmologist must remember that although the cause of blindness cannot be eliminated there may still be much to do. S/He also has a vital role to ensure the child gains early access to the services for the visually-impaired. Registration as blind or partially sighted, if relevant, is recommended because in general terms the child not registered is less likely to receive adequate support. Considered this way, the clinician can encourage parents that registration is not the end of the road, but a positive act to ensure support.

7. STAFF ISSUES
The ophthalmic screening and management of this potentially blinding condition requires a high degree of expertise. This is particularly relevant to treatment and its timing, as it is clinical assessment which poses the greater challenge rather than the treatment process. Mindful of the difficult times parents of a preterm baby have already been through and that the responsibility for care of the baby is with the staff of the neonatal unit, communication and liaison skills are also critical. The last must take into account the multidisciplinary nature of management, and the need sometimes to make arrangements between hospital and community.

The skills for effective and sensitive ROP management come with practice, and are only feasibly acquired by an ophthalmologist. Because the number of babies requiring treatment is small it is strongly recommended that one or two consultants in each area (with geographical not health authority connotations) should take a specific interest and gain the appropriate expertise and supervise training. Bearing in mind holidays, study leave and other absences provision for cover, possibly inter-district, should be borne in mind.
8. REFERENCES


APPENDICES

APPENDIX 1

International Classification Of Retinopathy Of Prematurity\textsuperscript{31,35}

SEVERITY BY STAGE

Stage 1 Demarcation line
This white line, lying within the plane of the retina and separating avascular from vascular retinal regions.

Stage 2 Ridge
The line of stage 1 has increased in volume to extend out of the plane of the retina. Isolated vascular tufts may be seen posterior to the ridge at this stage.

Stage 3 Ridge with extraretinal fibrovascular proliferation
This may be:
- continuous with the posterior edge of the ridge
- posterior, but disconnected, from the ridge
- into the vitreous

Stage 4 Retinal detachment – subtotal
- extrafoveal
- involving the fovea

Stage 5 Retinal detachment – total

Funnel:

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‘Plus’ disease
‘Plus’ disease is an indicator of activity. In order of severity ‘plus’ signs include:
- engorgement and tortuosity of the posterior pole retinal vessels
- iris vessel engorgement
- pupil rigidity
- vitreous haze
LOCATION BY ZONE
Retinal blood vessels grow out from the optic disc, and the zonal arrangement reflects this pattern of vascular development. Neural organisation of the retina, being centred on the fovea, is eccentric to the vascular pattern. The zones are centred on the optic disc but zone 3 is crescentic being widest in the temporal retina and absent nasally.

Zone 1
Extends from the optic disc to twice the disc-foveal distance – a radius of 30°.

Zone 2
Extends from the periphery of the nasal retina (ora serrata) in a circle around the anatomical equator. In the temporal retina, in the absence of an anatomical landmark, zone 3 cannot be identified precisely, and it can only be known to be entered with certainty when the nasal retina is fully vascularised.

Zone 3
Anterior to zone 2, is present temporally, inferiorly and superiorly, but not in the nasal retina.

Extent Of ROP
This is recorded as clock hours in each eye in the appropriate zone.

Other Clinical Signs
It is also important to take record at each examination, changes in the cornea, anterior chamber, iris, pupil, lens and vitreous.

Regressed Retinopathy of Prematurity
The vitreoretinal sequelae of regressed early phase ROP, while falling outside the boundaries of ROP classification, also need to be described. These fall into two broad categories: vascular and retinal changes involving the posterior and peripheral retina.
Peripheral Changes

Vascular
- Failure to vascularise peripheral retina
- Abnormal, nondichotomous branching
- Vascular arcades with circumferential interconnections
- Telangiectatic vessels

Retinal
- Pigmentary changes
- Vitreoretinal interface changes
- Thin retina
- Peripheral folds
- Vitreous membranes with or without retinal attachment
- Lattice-like degeneration
- Retinal breaks
- Traction/rhegmatogenous detachment

Posterior Changes

Vascular
- Tortuosity
- Straightening of vessels in temporal arcade
- Decrease in angle of insertion of major temporal arcade

Retinal
- Pigmentary changes
- Distortion and ectopic of macula
- Stretching and folding of retina in macular region leading towards the periphery
- Vitreoretinal membrane
- Dragging of retina over disc
- Traction/rhegmatogenous retinal detachment
APPENDIX 2

Information for all parents
(ideally included as part of the general information to all parents of preterm babies admitted to neonatal units)

All babies with birthweights under 1500g or who have been born 9 weeks or more premature have their eyes examined routinely at least once. This routine examination is called screening. We screen because very small babies are at risk of developing an eye condition called retinopathy of prematurity (ROP), also sometimes known by its old name of retrolental fibroplasias.

The main cause of ROP is prematurity itself, so the more prematurely the birth occurs the greater the risk of ROP occurring. The amount of oxygen treatment required and the baby’s general condition may also influence whether ROP develops or becomes severe. However, some premature babies who have no serious illnesses still develop ROP, while others who have been very poorly do not.

ROP affects the developing blood vessels of the retina which lines the inside of the back of the eye. Mild degrees of ROP are very common and, in these babies, recovery is complete.

The purpose of screening is to diagnose babies who develop severe ROP so that they can be treated effectively. Should this happen to your baby the situation will be fully discussed with you.

Examinations do not need to start until a few weeks after birth and for most babies these are all completed before s/he is discharged home. Sometimes an examination may be necessary after you take your baby home, and we will arrange to see your baby as an outpatient. It is very important that you keep this appointment.

ROP is very common and in most babies is mild, settling completely without treatment and does not therefore affect vision. For those few babies who do require treatment this is usually successful.

Please do not hesitate to ask if you need any further information.
APPENDIX 3

Information for parents of babies with ROP which might become severe
(to be given only to parents of babies with severe ROP, or on request)

All babies with birthweights under 1500g or who have been born 9 weeks or more premature have
their eyes examined routinely at least once. This routine examination is called screening. We
screen because very small babies are at risk of developing an eye condition called retinopathy of
prematurity (ROP), also sometimes known by its old name of retrolental fibroplasias.

The main cause of ROP is prematurity itself, so the more prematurely the birth occurs the greater
the risk of ROP occurring. The amount of oxygen treatment required and the baby’s general
condition may also influence whether ROP develops or becomes severe. However, some premature
babies who have no serious illnesses still develop ROP, while others who have been very poorly do
not.

ROP affects the developing blood vessels of the retina which lines the inside of the back of the eye.
Mild degrees of ROP (stages 1 and 2) are very common and in these babies recovery is complete.
The purpose of screening is to diagnosis those babies who develop more severe ROP (stage 3) as
this can affect vision, so that it can be treated effectively.

Unfortunately your baby has now developed severe retinopathy of prematurity. The purpose of this
information is to let you know that although your baby has a serious eye condition, treatment is
available.

In ROP the tips of the retinal blood vessels grow abnormally and in severe cases they grow into the
cavity of the eye and form scar tissue. At first this scar tissue is at the very edge of the retina and so
does not affect vision. Should scarring extend, it may distort the tissues inside the eye and cause
reduced vision. Once scar tissue has affect vision there is little hope of successful treatment. So,
screening must take place to diagnosis severe ROP before serious damage has occurred.

Treatment of severe ROP is by cryotherapy or laser therapy to the retina, and both forms of
treatment have the same end result.

- Cryotherapy freezes and retina by applying a pencil-sized probe against the outer surface of
the eye.
- Laser treatment involves shining a laser light through the pupil of the eye onto the retina.

Although treatment is successful in many babies, sometimes success is only partial.

All babies who have required treatment will need to be examined from time to time over a period of
a few years. Some will become short-sighted and require glasses, and some children develop
squints.

The full details of ROP and its treatment will be personally discussed with you.
APPENDIX 4

Working Party Membership

Working Party 1990
Professor Alistair Fielder, College of Ophthalmologists+
Professor Alex Garner, College of Ophthalmologists
Mr Stewart Johnston, College of Ophthalmologists
Professor Malcolm Levene, British Association for Perinatal Medicine+
Dr Janet Rennie, British Association for Perinatal Medicine
Mr Ed Schulenburg, College of Ophthalmologists

+Co-chair

Working party 1994
Mr David Clark FRCS, FRCOphth, Consultant Ophthalmologist, Walton Hospital, Liverpool*
Professor Alistair Fielder FRCP, FRCS, FRCOphth, Professor of Ophthalmology, University of Birmingham Medical School, Birmingham*
Dr Neil Marlow DM, MRCP, Senior Lecturer in Child Health, University of Bristol Medical School, Bristol **
Mr W Ed Schulenburg FRCS, FRCOphth, Consultant Ophthalmologist, Western Eye Hospital, London*
Dr A Michael Weindling MD, FRCP, Reader in Child Health, University of Liverpool Medical School, Liverpool**
Dr Andrew Wilkinson FRCP, Reader in Paediatrics, University of Oxford Medical School, Oxford**

* Royal College of Ophthalmologists
** British Association for Perinatal Medicine