Retinopathy of prematurity (ROP) is a developmental abnormality of the retina and vitreous in preterm infants which involves disordered vascularisation, cellular maturation and cellular differentiation. It is an important cause of visual impairment and the outcome can be improved if the disorder is detected by screening which allows appropriate treatment and follow-up (RCO/BAPM 1995, Schaffer et al 1992 and Watts 1992). This guideline for screening to detect ROP and its treatment, is based on the Guidelines for Care around preterm birth (NHMRC 1996).

Incidence and risk factors.

- Extremely preterm (< 28 weeks) - 61%, with severe disease (stage 3 or above) in 21%.
- Very preterm (includes all < 32 weeks) - 33%, with severe disease in 5-9%.
- In those that develop ROP, it is usually first detected at 30-45 weeks Postmenstrual age (PMA) and reaches stage 3 at a mean PMA of 37 (range 32-50) weeks.
- If infants have been screened and ROP does not appear until after 36 weeks PMA, it is unlikely to be severe.

Consequences of disease

In some infants this proliferative retinopathy progresses to inflammatory, haemorrhagic, and ultimately fibrotic retinal scarring and detachment, which may lead to blindness (Schaffer et al 1992, Watts 1992). There are four stages of acute ROP (see Appendix) and adverse outcome is associated with development of stage 3 and 4.

Diagnosis

The aims of screening for ROP are to identify ROP which has the potential to reach stage 3; and severe (stage 3) ROP which may require treatment.

Screening for ROP should:

- be carried out on all infants born at less than 32 weeks (or with a birthweight of less than 1500 grams);
- commence in all infants at 4-6 weeks postnatal age (and those born at less than 28 weeks should be examined by 32 weeks postmenstrual age); and
- be repeated at least every two weeks until vascularisation has progressed into the outer retina (zone 3).
- For infants due for transfer or discharge home, it is important to ensure that the screening process is completed. Regardless of ROP, all preterm infants need increased surveillance of visual function during childhood.

Examination technique

Eye drops to dilate the pupils and provide local anaesthesia should be given at least 30 minutes before the examination. The precise type of medication used might vary but may include single drops of amethocaine 0.5%, tropicamide 0.5%, cyclopentolate 0.25% and phenylephrine 2.5% instilled into each eye. Examination should take place in the usual environment of the infant (eg humidicrib) with minimal handling and disturbance of the infant and maintenance of current monitoring and therapy (eg oxygen).
Record keeping

Using the International Classification of ROP (CCRP 1987, RCO/BAPM 1995), the following signs should be recorded (for details see Appendix C):

- severity by stage
- location by zone
- extent in clock hours
- ‘plus’ disease
- other clinical signs such as: 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.

Interventions

When to intervene

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. Nevertheless, this group still requires follow-up for less severe visual problems. The threshold for treatment is therefore defined as stage 3 ROP, involving five or more contiguous, or eight or more cumulative, clock hours in the presence of congestion of the posterior pole vessels - ‘plus’ disease. The latter is an indicator of activity and, 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 there is vitreous haze and sometimes when there are iris changes, the results of treatment are poor. Treatment should be undertaken as soon as possible, ideally within 2-3 days of the identification of threshold disease. The aim of treatment is to remove the stimulus for vessel growth, ie ablate the peripheral avascular retina.

Intervention

Retinal ablation is effective treatment. Meta-analysis of the results of two randomised controlled trials with consistent results shows that when ROP has reached threshold (stage 3+), cryotherapy 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 can also be performed using laser. There is some evidence to suggest that diode is preferable to argon laser (ref).

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.

Preparing the baby

Preparation of the baby is essential to ensure minimal harm and close supervision. The eye should be prepared by dilating the pupils fully, bearing in mind the potential systemic toxicity of these topical agents. Subconjunctival or retrobulbar anaesthetics must not be used.
Monitoring during treatment

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 essential. Many believe that the optimal condition is for the baby to be intubated and given artificial ventilation. Experienced staff must be in attendance throughout and cardiorespiratory monitoring which must be continued until the baby has fully recovered. Staff should be aware of the systemic (bradycardia, cyanosis) and local (oedema, haemorrhage, burns) complications of therapy that might arise in the subsequent hours and days, and monitor for them.

Counselling parents

For parents of all babies at risk

Written information about the need to screen for ROP should be provided as part of the general information provided by the neonatal unit for parents of premature infants. It is important to emphasise that mild ROP is very common and spontaneously resolves without adverse sequelae in the vast majority.

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 discuss 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 a senior neonatal nurse, is present so that post-interview queries can be dealt with. Severe ROP often occurs just when parents are beginning to relax for the first time after stressful weeks of uncertainty.

Parents should be kept 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.

For parents of infants with advanced ROP

The need to keep in close contact with the family cannot be overemphasised. Generally, parents can accept with time that not all disorders can be successfully treated, but they cannot accept lack of interest or care.

Follow-up

All infants with ROP of any stage require a follow-up examination the first 6 months of life and then at intervals as dictated by the findings. Infants with stage 3 ROP and those who have received therapy require particularly close surveillance in the preschool years as the incidence of strabismus and other problems is particularly high in this group.

Parents of infants with severe visual impairment need support and advice about treatment options. Referral to specific support groups for the visually impaired, such as State/Territory societies or institutes for the blind, should be offered.

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