Retinopathy of Prematurity: A Practical Clinical Approach

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Objectives

After completing this article, readers should be able to:

1. Describe the forms that should be used to record all retinopathy of prematurity (ROP) screening examinations.
2. Characterize the roles of ophthalmologists and neonatologists in developing local screening policies.
3. Delineate which preterm infants require screening for ROP and when the first screening examination should occur.
4. Describe the procedure for laser therapy to treat ROP and postsurgical follow-up.

Introduction

The neonatologist and neonatal intensive care unit (NICU) staff have a serious responsibility to minimize vision loss from retinopathy of prematurity (ROP). It requires a team effort throughout the hospitalization to reduce the incidence and severity of ROP and particularly conscientious tracking of patients during the phase of active disease to detect and treat the few cases that become severe enough to threaten retinal detachment. It is particularly frustrating in this most visual of diseases that the pathology occurs inside the eye where the threat is not visible and at the particular time when the infants are recovering from so many other insults and are nearly ready for home!

Reducing the Risk of ROP

Among infants of similar gestations, those who have the more unstable hospital course have the higher risk of serious ROP. It is important to remember that interventions that prolong pregnancy and reduce intraventricular hemorrhage, pneumothorax, serious sepsis, and chronic lung disease all reduce the risk of vision-threatening ROP. To date it is impossible to prevent all morbidities, but each year brings increased knowledge about how to reduce their incidence and hopefully the incidence of severe ROP.

Creating a Screening and Tracking Program

An effective ROP screening program requires the cooperative efforts of the neonatal team, screening ophthalmologists, and discharge coordinators. Unlike many screening programs in which the “screening test” is inexpensive, the time and effort of ophthalmologists is considerable in this program, and rarely are their consulting fees paid. Participating in ROP screening is an effort of dedication to preserve vision. The disruption to NICU feeding and care routines is not trivial, and careful coordination of the entire plan can minimize general frustration.

A limited number of ophthalmologists who have a sincere interest in ROP should be recruited. More than one is needed to have coverage during vacations/illness, but a panel of 12 might leave each with insufficient ongoing experience to maintain optimal skills.

Selection of Infants for Screening

Ophthalmologists and neonatologists should agree jointly upon which infants will be screened and when in their hospitalization examinations will be initiated. Formal screening statements have been published in the United States, Canada, and England, and they are reasonably similar (Table). A written local policy should be developed from these recommendations that covers the topics described in this section. It helps to establish routine

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orders for administration of dilating drops, sterilization procedures for lid specula and scleral depressors, and the relationship of examinations to feedings. A regular time and examination day of the week is extremely helpful when several examinations must be performed each week. Most ophthalmologists prefer a preprinted consultation form for ROP screening that documents the infant’s gestation, birthweight, and the request for consultation. Providing blank sketches of the retinal zones and disk, as suggested by the ICROP, facilitates the quick drawing of retinal findings. Summary boxes that permit easy (and legible) conclusions are also a great benefit. An example of a useful screening form is provided in Figure 1. The critical elements are the description of the ROP or degree of immaturity using ICROP and the recommendation for repeat examination.

The policy must address who to screen and when, which are key elements of the various formal statements (Table). Applying these statements, however, requires some interpretation, particularly the statement that “... those infants over 1,500 grams with an unstable clinical course felt to be at high risk by their attending ... should have an ... examination to detect ROP” from the United States statement. There are no recommendations about who these bigger preterm infants are, but generally those who have required ventilation, who have been hypotensive, or who have received prolonged oxygen are included by most NICUs. Infants of 37 weeks’ gestation and greater do not need to be screened for ROP, even if they do have an unstable course.

**Timing of Screening**

Determining when to perform the initial screening examination is a balance of reducing unnecessary examinations and not missing threshold ROP near its onset. Because ROP is rarely if ever observed in the first 4 weeks following delivery or before 31 weeks postmenstrual age (PMA), the first examination can be delayed until 31 weeks PMA in the smallest infants younger than 27 weeks’ gestation and until 4 weeks chronologic age in the more mature infants of 28 weeks’ gestation and greater who will be examined. The neonatal team should make the request for consultation and examination and verify that the infant is sufficiently stable to tolerate the examination. If a particularly critical time frame is reached in an unstable infant, rather than miss the opportunity to treat threshold ROP, a very limited examination of minimal stress can rule out plus disease, and therefore, the need for surgery. A more complete examination can be performed the following week, when the infant has stabilized.

The recommendation for the timing of a repeat examination is determined by the ophthalmologist based on the findings of the first examination. However, generally agreed-upon principles should be placed in the policy. This contributes to consistency and the usual medical double-checking of discharge plans. Normally, threshold ROP requires urgent consultation from a surgeon who could perform laser or cryotherapy (within 2 to 3 days) if indicated. Prethreshold ROP requires a return visit within 1 week; milder ROP is evaluated at 2-week intervals unless it has progressed into zone III. Zone III ROP is very low risk and can be followed at longer intervals. However, it is important to remember that very immature infants are extremely unlikely to have either zone III vessels or a “mature retina” on a first examination, and the examination should be repeated at least once if this unusual event is recorded on the first examination.

**Communication**

Communication is critical in a screening program that involves bedside nurses, families, neonatologists, ophthalmologists, retinal surgeons, house staff, and discharge coordinators. The written policy and tracking of

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*PMA = postmenstrual age, which is the gestational age plus the chronologic age since birth in weeks.

Figure 1. ROP Screening Form from the University of Rochester.
infants for the first examination should be assigned to appropriate personnel, with back-up for vacations. Neonatologists must assist in determining which preterm infants older than 28 weeks’ gestation have been “medically unstable.” Ophthalmologists must clearly record the retinal findings using ICROP, preferably on a consultation form with retinal sketches. The assistance of a limited number of special helpers for the ophthalmologist during examinations in a high-volume NICU is particularly valuable. They can facilitate the preparation of paperwork, completion of forms, tracking of the tiny, expensive instruments, and communication with the bedside nurses and staff.

Families are normally well known by the neonatal team members, who can communicate the results of the eye examination. Most units hand out general information pamphlets to families around the time of the first screening examination, which helps to smooth communications. Use of a standardized form helps the team to provide predictions to the family. One helpful practical communication device is to tell the parents that repeat examinations every 2 weeks represents the usual course, and that they can begin to be concerned when the repeat examinations are scheduled in 1 week or less. When the ROP becomes concerning (prethreshold and especially threshold), the ophthalmologist or another clinician very familiar with ROP should try to communicate directly with the family. Threshold ROP requires timely consultation from a surgeon who can perform laser or cryotherapy. The examiner in these cases must describe the gravity of the situation to the consultant as soon as feasible, which is usually by phone the same day because arrangements for treatment to be completed within 72 hours can be challenging.

An infant can be treated with laser or cryotherapy in either the operating room with anesthesia support or in the NICU with neonatology support. The procedure on the eye is moderately painful, but narcotic analgesia is usually sufficient, sometimes aided by local anesthesia. Protection of the airway and monitoring of ventilation and oxygenation is, as always, critical for the procedure. Pressure on the eyeball occasionally can trigger the oculocardiac reflex, resulting in bradycardia. Atropine is useful for cases when this may be a problem. The pupils must be extremely well dilated, and because threshold ROP impairs iris dilatation, the surgeon will request stronger dilating drops than are used for screening examinations. If these are part of the unit policy, time and confusion can be spared by having the appropriate drugs routinely available.

Threshold ROP usually develops when other medical problems are finally resolving, so reintubation for this procedure is a particularly disturbing event for parents.

**Discharging With ROP**

ROP is becoming an increasing problem for discharge planning. Infants are being back-transferred or discharged home earlier and more frequently on oxygen. As can be seen from Figure 2, threshold ROP commonly (about 50% of cases) occurs at 36 to 39 weeks and earlier discharge dates from the NICU, the high risk circumstance of an infant going to threshold following discharge home is increasing. Modified from Palmer, et al. Ophthalmology. 1991;98:1628–1640.
Clinical Care Following Laser/Cryotherapy

Fortunately, ROP regresses (improves) after treatment in most infants. In the STOP-ROP study, 82% of the treated eyes had good anatomic outcomes (no retinal detachment, folds, or dragged disks). Other than the immediate postoperative period, when increased apnea and local swelling may be observed, recovery is usually uneventful. In some eyes, however, retinal detachments do occur. Rescue efforts to reattach the retina or stop the progression of the detachment are disappointing. Scleral buckling occasionally can permit the somewhat shrunken retina to reattach and is offered in some cases of stage 4 ROP. Vitrectomy for stage 5 total retinal detachments is considered a heroic attempt at retinal rescue, and sometimes light/dark perception or ambulatory vision can be achieved. However, these eyes very rarely have been reported to have measurable visual acuity. It is important that families who choose to try such surgery have realistic expectations about the outcome. The neonatal team can be helpful in discussions accompanying these decisions.

Long-term Outcomes of ROP

Planning is needed for lifetime follow-up of ROP, and counseling begins in the neonatal period. All preterm infants have a higher risk of crossed-eye and refractive errors and are usually, therefore, evaluated by an ophthalmologist at 6 to 12 months. If an infant’s ROP has been less than prethreshold, this follow-up plan is still appropriate. If the ROP reached a severity of prethreshold but no worse, the outcomes are generally the same, and the usual preterm infant follow-up schedule is appropriate.

If threshold ROP has occurred, however, there are high rates of severe refractive error (usually extremely high myopia/near-sightedness). These infants should be evaluated by a pediatric ophthalmologist or similarly interested general ophthalmologist at around 3 months of corrected age to determine their refractive error and provide corrective lenses and amblyopia treatment as indicated. Without these, formed vision at a distance of more than 2 or 3 inches is denied an infant who might otherwise be acquiring visual data about his or her environment.

At least annual re-examinations are recommended for any infants who have residual peripheral retinal scars because of their unknown prognosis. The oldest adults who had ROP are now in their late 50s, and some have experienced late retinal detachments in their 20s and 30s after escaping vision loss in the newborn period. The oldest preterm infants treated with cryotherapy or laser therapy are now teenagers, and we have yet to learn what those treatment scars will do in the long term. Late retinal detachments from a retinal tear or hole are more easily treated than the massive retinal detachments of the neonatal period from ROP. All children who had threshold ROP should establish a lifelong follow-up arrangement with ophthalmologists for the monitoring of residual retinal scars (cicatrix).

Infants who lose vision despite treatment are also at special risk and have a higher incidence of functional impairments. Special education for families and infants who have visual impairment makes significant differences in learning skills and life function. Infants who have retinal detachments used to face long months, or sometimes years when families would not believe their child’s blindness. With early communication and information about ROP as it develops, this seems to be less of a problem today. Affected families should be referred to early intervention programs as soon as the diagnosis is
known. The British publication on ROP management is particularly interesting and broad-based reading (Early Hum Dev. 1996;46:239–258). Many state and national services are available, and teaching families how to teach their infants to experience the world with their remaining senses is the key to optimizing outcomes as the children grow.

Suggested Reading


NeoReviews Quiz

6. You are teaching medical students about the epidemiology of ROP. Of the following, the most accurate statement about ROP is that:
   A. Black infants are more susceptible than white infants to threshold ROP.
   B. ROP is seen rarely before 31 weeks of postmenstrual age.
   C. The incidence of ROP specific for gestational age has increased over the last decade.
   D. The primary cause of ROP is exposure to supplemental oxygen.
   E. The zone of ROP changes during evolution of the disease in an infant.

7. You are counseling parents of a preterm infant about the surveillance and treatment of ROP. Of the following, the most accurate statement is that:
   A. Pain control during cryotherapy usually is achieved with general anesthesia.
   B. The initial eye examination should be performed within 2 weeks after birth.
   C. The risk of disease progression decreases during convalescence from surgery.
   D. Threshold retinopathy impairs dilation of the iris.
   E. Unstable near-term infants should undergo eye examination for ROP.

8. An ophthalmologist examines a preterm infant as a part of surveillance for ROP. Based on the findings of this examination, she recommends a repeat eye examination 1 week later. Of the following, the most likely retinal finding consistent with this recommendation is:
   A. Mature retina.
   B. Prethreshold ROP.
   C. Stage 2 ROP.
   D. Threshold ROP.
   E. Zone III ROP.