

Screening for Retinopathy of Prematurity and Risk Management

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- I. Complete Knowledge of Retinopathy of Prematurity (ROP)
 - A. Able to easily recognize all stages of disease, including normal immature retina and variants of disease
 - 1. Immature developing normal retina
 - a. Normal choroidal pattern: Not fully vascularized
 - b. Junction of vascularized and non-vascularized retina
 - 2. Stage 1: Demarcation line
 - 3. Stage 2: Ridge
 - 4. Stage 3: Neovascular (NV) ridge
 - a. Brushborder NV ridge
 - b. Elevated NV ridge
 - c. Flat NV ridge
 - d. Regressed, popcorn NV
 - 5. Stage 4: Partial retinal detachment (RD)
 - a. Stage 4A retinal detachment: Fovea attached
 - b. Stage 4B retinal detachment: Fovea detached
 - 6. Stage 5: Total retinal detachment
 - a. Open funnel total retinal detachment
 - b. Closed funnel total retinal detachment ⊗ leukocoria
 - 7. Plus disease: Vascular dilation and tortuosity
 - a. Normal infant retinal vessels (thin, wire-like, no tortuosity)
 - b. Mild plus disease
 - c. Moderate plus disease
 - d. Marked plus disease
 - B. Zones of disease
 - 1. Zone I: Circle with a radius of 30 degrees centered on optic nerve head (any part of ridge within view of 28 diopter indirect lens)
 - 2. Zone II: From Zone I temporally to the temporal equator and to the ora serrata nasally
 - 3. Zone III: From Zone II temporally to the ora serrata temporally
 - C. Clock hour involvement: 0 through 12 clock hours involved (for use with Stages 1 through 4)
 - D. Difference in view of ROP from reference pictures
 - 1. American Academy of Ophthalmology (AAO) standard Kowa camera views of disease states → High magnification
 - 2. Standard indirect ophthalmoscope view with a 28 diopter lens → Standard (medium) magnification
 - 3. Retcam (130 degree) ROP lens view → Lower magnification, wide field view

Pediatric Retinal Diseases

E. Factors hampering view of disease

1. Baby looking away
 - a. Alphonso lid speculum useful
 - b. Flynn scleral depressor useful
2. Poor dilation
 - a. ? Due to baby squeezing dilating drops out
 - b. ? Due to advanced plus disease not allowing for good dilation
3. Mildly hazy, normal premature cornea: Naturally occurring early in life in some very premature infants
4. Persistent tunica vasculosa lentis (TVL) in premature babies
 - a. Due naturally to early postconceptional age (PCA)
 - b. Due to abnormal blood vessel activity (plus disease)
5. Flare in the vitreous cavity: Due to breakdown of the blood-ocular barrier, usually associated with advanced plus disease
6. Vitreous hemorrhage
 - a. Due to advanced ROP
 - b. Due to birth trauma

F. Pre-threshold disease

1. Any Zone I ROP
2. Zone II ROP, Stage 3, without plus disease
3. Zone II ROP, Stage 2, with plus disease
4. Zone II ROP, Stage 3 (less than threshold clock hours), with plus disease

G. Threshold disease

1. Zone I or II ROP, Stage 3 (5 contiguous clock hours), with plus disease

2. Zone I or II ROP, Stage 3 (8 cumulative clock hours), with plus disease
3. With posterior disease (Zone I, or border Zone I/Zone II), be aware of *flat* Stage 3 that may be difficult to detect

H. Retinal detachment (Stage 4)

1. Early
 - a. Evolving traction on NV
 - b. Dragging of retinal vessels/ macula
2. Late
 - a. Loss of choroidal vascular pattern on indirect ophthalmoscopy viewing
 - b. Peripheral "white retina" sign due to double thickness dragged retina

I. Danger signs associated with aggressive disease

1. Very low birth weight (BW) baby, < 1000 grams (g)
2. Early onset of disease
3. Posterior disease
4. Rapid tempo of disease progression
5. Marked vascular activity with a significant TVL

II. Screening Recommendations for Babies at Risk for ROP

- A. Initial eye exam at 31 weeks postconceptional age or 4 weeks chronological age (whichever is later)
 1. BW < 1500 g, or
 2. Gestational age (GA) < 28 weeks, or
 3. Selected babies with a BW between 1500 g and 2000 g with an unstable clinical course
- B. Adequate pupillary dilation with indirect ophthalmoscopy
- C. At least 2 exams to detect ROP

D. Exam timing should allow for sufficient time to treat plus extra time if transferring the baby is necessary

E. Follow-up examinations: Based on findings at the previous exam

1. Pre-threshold disease → 1 week follow-up
 - a. Zone I ROP (not yet threshold)
 - b. Zone II ROP, Stage 3, without plus disease
 - c. Zone II ROP, Stage 2, with plus disease
 - d. Zone II ROP, Stage 3 (less than threshold clock hours), with plus disease
2. Zone I, without ROP → 1 to 2 weeks follow-up
3. Less severe ROP in Zone II → 2 weeks follow-up
4. Immature, Zone II, no plus disease → 2 to 3 weeks follow-up

F. Acute phase ROP screening can be discontinued when the risk of ROP visual loss is minimal or passed when:

1. Fully mature retinal vasculature (to the ora serrata nasally or within 1 disc diameter from the ora serrata temporally) is present
2. 45 weeks PCA is reached without the infant having previously developed pre-threshold ROP or worse
3. Progression of normal retinal vessels into Zone III, *without* prior Zone II ROP

III. Threshold Disease

A. Definition:

1. > 5 contiguous clock hours of Stage 3, with plus disease
2. 8 total clock hours of Stage 3, with plus disease

B. Treat within 72 hours

C. Diode laser: Complications include:

1. Risk of progression of disease
2. Cataract
3. Vitreous hemorrhage
4. Retinal detachment
5. Anterior segment ischemia
6. Blindness

D. Follow-up 1 week post laser treatment

E. If plus disease persists at 2 weeks post laser treatment, consider repeating laser treatment.

F. Look for development of retinal detachment (occurs most commonly at 38 to 45 weeks PCA).

G. Once RD develops, it can progress rapidly (ie, over a few weeks to advanced stages).

H. If RD develops, then treat (with laser vs scleral buckle vs vitrectomy) or refer promptly.

IV. Medicolegal Issues

A. From the outset of an interaction with a patient, keep the parents informed of the nature and possible severity (blindness) of ROP. Ideally give a prepared handout from the AAO or Ophthalmic Mutual Insurance Company (OMIC).

B. Keep parents informed of progression of the condition during the course of follow-up and treatment.

C. Emphasize to parents the need for prompt and close follow-up after hospitalization, and point out the risk of irreversible blindness if this is not adhered to (ie, missed appointments).

D. Document all findings and discussions completely and well in the patient's record.

Pediatric Retinal Diseases

- E. Explain to the parents from the beginning that even with proper and prompt diagnosis and treatment, there is a risk of blindness and possible need for further surgery.
- F. Have documentation and agreement between the neonatal intensive care unit (NICU) and ophthalmologist that states that the coordination of the initial screening exam and follow-up exams is the responsibility of the hospital while the infant is in the hospital.
- G. If the infant is too sick for an eye exam, then this should be documented by the neonatologist.
- H. If an infant is transferred from the NICU before retinal maturation is complete, then it is the transferring primary physician who is responsible for arranging/ communicating and documenting the need for proper and timely ophthalmology follow-up exams with the new primary physician.
- I. If the infant is being discharged from the NICU before retinal maturation is complete, it should be communicated to the parents and documented with their signature that they clearly understand the risk of irreversible blindness if they do not strictly adhere to proper and timely follow-up.

References

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