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**ROP Safety Net Toolkit**

**Anne M. Menke, RN, PhD**

**Reviewed by**

**Daniel M. Berinstein, MD; Denise R. Chamblee, MD;**

**Robert S. Gold, MD; and Christie L. Morse MD**

**OMIC policyholders who provide care must comply with the ROP Safety Net.**

OMIC’s ROP Safety Net is based on our claims experience. It is designed to address the causes of ROP lawsuits in order to protect the infant and the ophthalmologist. The ROP Safety Net Toolkit contains sample protocols, which may need to be customized, and refers to ROP clinical care guidelines. These protocols and guidelines are recommendations and do not constitute the standard of care. Ophthalmologists should use their professional judgment in determining the applicability of a given recommendation to their particular patients and practice situation.

The Toolkit does not provide legal advice. Consult an attorney if legal advice is desired or needed. Information contained here is not intended to be a modification of the terms and conditions of the OMIC professional and limited office premises liability insurance policy. Please refer to the OMIC policy for these terms and conditions.

**Version 8/10/18**

# **Procedure 2a. ROP exam in hospital**

**Use the hyperlinks to see tables and forms. To go back to where you were in the document using a PC, press Alt+left arrow.**

**ROP exam in hospital principles**

1. The hospital has a treating ophthalmologist with staff privileges, or has a transfer agreement in place with a hospital that can accept the transfer and provide ROP treatment within 72 hours.
2. The hospital admits infants from the outpatient setting who need ROP treatment, or has a transfer agreement in place with a hospital that does admit infants and can provide ROP treatment within 72 hours.
3. The hospital has an ROP coordinator (H-ROPC) who:
	1. Is familiar with and understands the ROP Screening Policy Statement (PS)[[1]](#footnote-1) and the Tables in the ROP toolkit that are based upon it, and is able to use the Tables to review and clarify the appropriateness of follow-up and treatment intervals, and coordinate discharge or transfer.
	2. Keeps the Master Hospital [ROP Tracking List](#_ROP_Tracking_List) of hospitalized infants who need ROP care, and sends the office ROPC (O-ROPC) a copy of it.
	3. Reviews the Hospital ROP Tracking List with the O-ROPC **at least once a week.** The ROPCs compare the current list with the list from the prior week. The H-ROPC notifies the ophthalmologist and neonatologist of all missed, cancelled, or rescheduled ROP exams.
4. The ophthalmologist:
	1. Has an O-ROPC who works with the H-ROPC to track infants needing ROP care.
	2. Has sufficient knowledge and experience to identify accurately the location and sequential retinal changes of ROP after pupillary dilation using binocular indirect ophthalmoscopy with a lid speculum and scleral depression as needed (PS #2).
	3. Uses the International Classification of Retinopathy of Prematurity ([ICROP](#_ICROP._Synopsis_of)) Revisitedto classify, diagram, and record the retinal findings (PS #2).
	4. Knows and understands treatment criteria [[Table 4. When to treat](#_Table_4._)].

**Hospital ROP exam procedure**

**Use the hyperlinks to see tables and forms. To go back to where you were in the document using a PC, press Alt+left arrow.**

1. The neonatologist identifies new infants who meet screening criteria [[Table 1. Who to screen](#_Table_1._Which_1)] and indicates the approximate date of the initial ROP exam [[Table 2. When to start](#_Table_2._When)].
2. The neonatologist instructs the H-ROPC to add the infant’s name and date of the initial ROP exam to the Hospital [ROP Tracking List](#_Tracking_List).
3. The H-ROPC contacts the O-ROPC of the screening ophthalmologist to schedule the initial ROP exam and begin tracking.
4. The H-ROPC or NICU nurse assists the ophthalmologist with the exam and:
	1. Reviews the list of infants to be examined that day, along with their medical records.
	2. Consults with the neonatologist to determine if any contraindications to the examination exist and:
		1. Notifies the O-ROPC of any infant who cannot undergo the scheduled eye exam.
		2. Reschedules the exam within the time interval indicated by the infant’s most recent eye exam.
		3. Contacts the neonatologist and ophthalmologist to determine the best course of action, and documents the discussion, if the infant cannot be examined within the indicated interval.
		4. Documents the notification and reason for not having the exam in the infant’s medical record.
		5. Notifies the parent of the delay and documents the discussion.
	3. Provides the following supplies:
		1. Sterile NICU eye tray with lid speculum and depressor
		2. Anesthetic eye drops
		3. Indirect ophthalmoscope (if ophthalmologist does not bring one)
		4. 20 and 28 diopter lenses
		5. Dilating eye drops
		6. Gloves
	4. Dilates the infants’ eyes at the time ordered by the ophthalmologist per the dilating protocol.
	5. Ensures that participants in the eye exam have washed their hands, and, if indicated, wear gloves to prevent eye irritation and infection.
	6. Secures the infant in a blanket, holds the infant during the exam, and provides a pacifier and/or oral sucrose for comfort.
	7. Monitors the infant for side effects associated with the dilating eye drops and exam.
	8. Documents the dilation, exam, and the infant’s condition during the exam.
	9. Cleans and sterilizes the equipment according to the manufacturer’s specifications to prevent eye irritation and infection.
5. The ophthalmologist performs a binocular indirect ophthalmoscopy (BIO) exam after pupillary dilation and documents the findings using [ICROP](#_ICROP._Synopsis_of)).
6. The ophthalmologist determines the timing of the next examination [[Table 3. Follow-up exams](#_Table_3.__1)].
	1. Current guidelines indicate a range of 1 to 3 weeks between examinations, depending upon the findings.
	2. Infants at high risk for ROP may need more frequent examinations.
	3. Infants treated with an anti-VEGF medication (i.e., Avastin or Lucentis) need to be monitored for a longer period of time.
7. The ophthalmologist writes an order for the next exam indicating the interval **and** approximate date (e.g., next eye exam in two weeks around 9/25/18) and:
	1. Notifies the ROPCs of the next exam interval and approximate date, and instructs them to update the Hospital ROP Tracking List.
8. The ophthalmologist screens for ROP until one of the following conditions has been met and documented:
	1. ***Per the Policy Statement, one exam is sufficient only if it unequivocally shows the retina to be fully vascularized in both eyes.***
	2. A treating ophthalmologist has verified that the treatment and follow-up examinations are complete.
	3. Both eyes have met the conclusion-of-acute-screening criteria based upon a BIO exam [[Table 5. When to stop](#_Table_5._)].
	4. The current ophthalmologist conducts and documents a transfer-of-care discussion with the ophthalmologist who will take over care.

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# **[Table 1. Which infants need an ROP screening examination](#Table_1)**

Infants meeting any of the following criteria need an exam:

* Birth weight of ≤ 1500 g (3 lbs., 4 oz.)
* Gestational age of 30 weeks or less (as defined by the attending neonatologist)
* Selected infants with a birth weight between 1500 and 2000 g (from 3 lbs., 4 oz. to 4lbs, 6 oz.) or gestational age of more than 30 weeks with an unstable clinical course, including those requiring cardiorespiratory support and who are believed by their attending pediatrician or neonatologist to be at high risk for ROP.

**REFERENCE: ROP Screening Policy Statement # 3**. Based on Recchia, Franco and Capone, Antonio, Contemporary Understanding and Management of Retinopathy of Prematurity, *Retina* 2004; 24:283-92.

# **[Table 2. When to start ROP screening](#Table_2)**

The onset of serious ROP correlates better with postmenstrual age (gestational age at birth plus chronological age) than with postnatal age. This protocol bases the initial eye examination on postmenstrual age and chronological age. The initial eye examination should be conducted:

* By 31 weeks postmenstrual age if gestational age < 27 weeks
* At 4 weeks chronological age if gestational age ≥ 27 weeks

**Age in weeks at initial exam**

|  |  |  |
| --- | --- | --- |
| **Gestational age at birth**  | **Postmenstrual age** | **Chronologic age** |
| 22a\* | 31 | 9 |
| 23a\* | 31 | 8 |
| 24\* | 31 | 7 |
| 25\* | 31 | 6 |
| 26 | 31 | 5 |
| 27 | 31 | 4 |
| 28 | 32 | 4 |
| 29 | 33 | 4 |
| 30 or more | 34 | 4 |
|  |  |  |

a This guideline should be considered tentative rather than evidence-based for 22-to-23-week infants owing to the small number of survivors in these gestational age categories.

**\*** Infants born before 25 weeks’ gestational age should be considered for earlier screening on the basis of severity of comorbidities (6 weeks’ chronological age, even if before 31 weeks’ postmenstrual age, to enable earlier identification and treatment of aggressive posterior ROP [a severe form of ROP that is characterized by rapid progression to advanced states in posterior ROP] that is more likely to occur in this extremely high-risk population).

**REFERENCE:** **ROP Screening Policy Statement #3.** Based upon Reynolds JD, Dobson V, Quinn GE, et al. CRYO-ROP and LIGHT-ROP Cooperative Groups. Evidence-Based Screening Criteria for Retinopathy of Prematurity: Natural History Data from the CRYO-ROP and LIGHT-ROP Studies. *Arch Ophthalmol.* 2002; 120: 1470-1476.

# **[Table 3. Follow-up schedule for ROP exams](#Table_3)**

The examining ophthalmologist should use retinal findings as classified by [ICROP](https://jamanetwork.com/journals/jamaophthalmology/fullarticle/417157) to determine the timing of the follow-up examinations.

* 1-week or less
	+ Immature vascularization in zone 1—no ROP
	+ Immature retina extends into posterior zone II, near the boundary of zone I
	+ Stage 1 or 2 ROP in zone I
	+ Stage 3 ROP in zone II
	+ The presence or suspected presence of aggressive posterior ROP
	+ Infants treated solely with anti-VEGF medications such as bevacizumab
* 3 to 7 days
	+ After treatment to ensure that there is no need for additional treatment in areas where ablative treatment was not complete.
* 1 to 2 weeks
	+ Immature vascularization in posterior zone II
	+ Stage 2 ROP in zone II
	+ Unequivocally regressing ROP in zone I
* 2 weeks
	+ Stage 1 ROP in zone II
	+ Immature vascularization in zone II—no ROP
	+ Unequivocally regressing ROP in zone II
* 2 to 3 weeks
	+ Stage 1 or 2 ROP in zone III
	+ Regressing ROP in zone III

**REFERENCE**: **ROP Screening Policy Statement #4**. Based on Reynolds JD, Dobson V, Quinn GE, et al. CRYO-ROP and LIGHT-ROP Cooperative Groups. Evidence-Based Screening Criteria for Retinopathy of Prematurity: Natural History Data from the CRYO-ROP and LIGHT-ROP Studies. *Arch Ophthalmol.* 2002; 120: 1470-1476.

# **Table 4. When to treat ROP**

* Treatment should be initiated for the following retinal findings:
	+ Zone I ROP: any stage with plus disease
	+ Zone I ROP: stage 3—no plus disease
	+ Zone II ROP: stage 2 or 3 with plus disease
* The presence of plus disease in zones I or II suggests that peripheral ablation, rather than observation, is appropriate.\*
	+ Plus disease is defined as abnormal dilatation and tortuosity of the posterior retinal blood vessels in 2 or more quadrants of the retina meeting or exceeding the degree of abnormality represented in reference photographs
* Consideration may be given to treatment of infants with zone I stage 3+ ROP with intravitreal injection of bevacizumab.#
	+ Bevacizumab is not approved by the US Food and Drug Administration for the treatment of ROP.
	+ Treatment should only be administered after obtaining detailed informed consent, because there remain unanswered questions involving dosage, timing, safety, visual outcomes, and other long-term effects.
	+ Infants treated with bevacizumab should be monitored weekly until retinal vascularization is complete.
	+ Longer follow-up is required because recurrence occurs considerably later (16 ± 4.6 weeks vs 6.2 ± 5.7 weeks) than after laser therapy.
* Special care must be used in determining the zone of disease.
	+ See page 992 of [ICROP](https://jamanetwork.com/journals/jamaophthalmology/fullarticle/417157) for specific examples of how to identify zone I and II disease by using a 28-diopter lens with binocular indirect ophthalmoscopy.
* The presence of plus disease rather than the number of clock hours of disease may be the determining factor in recommending ablative treatment.
* Treatment should generally be accomplished, when possible, within 72 hours of determination of treatable disease to minimize the risk of retinal detachment.
* Follow up is recommended in 3 to 7 days after treatment to ensure that there is no need for additional treatment in areas where ablative treatment was not complete.

**REFERENCE: ROP Screening Policy Statement #7 and #9 based upon:**

\* Early Treatment for Retinopathy of Prematurity Cooperative Group. Revised Indications for the Treatment of Retinopathy of Prematurity. Results of the Early Treatment for Retinopathy of Prematurity Randomized Trial. *Arch Ophthalmol.* 2003; 121:1684-1694.

* # Mintz-Hittner HA, Kennedy KA, Chuang AZ; BEAT-ROP Cooperative Group. Efficacy of intravitreal bevacizumab for stage 3+ retinopathy of prematurity. *N Engl J Med*. 2011; 364(7):603–615.

# **Table 5. When to stop ROP screening**

**Per the Policy Statement, one exam is sufficient only if it unequivocally shows the retina to be fully vascularized in both eyes.**

The conclusion of acute-retinal-screening examinations should be based on age and retinal ophthalmoscopic findings. Findings that suggest that examinations can be terminated include:

* Zone III retinal vascularization attained without previous zone I or II ROP
	+ If there is examiner doubt about the zone or if the PMA (postmenstrual age) is less than 35 weeks, confirmatory examinations may be warranted.
* Full retinal vascularization in close proximity to the ora serrata for 360°--that is, the normal distance found in mature retina between the end of vascularization and the ora serrata.
	+ **Per the Policy Statement, this criterion should be used when ROP is treated solely with anti-VEGF medication.**
* Postmenstrual age of 50 weeks and no prethreshold disease or worse ROP is present
	+ Prethreshold disease defined as:
		- Stage 3 ROP in zone II
		- Any ROP in zone I
* Regression of ROP (see [ICROP](#_Appendix_B._))
	+ Care must be taken to be sure that there is no abnormal vascular tissue present that is capable of reactivation and progression in zone II or III.

**REFERENCE: ROP Screening Policy Statement # 5.** Based upon Reynolds JD, Dobson V, Quinn GE, et al. CRYO-ROP and LIGHT-ROP Cooperative Groups. Evidence-Based Screening Criteria for Retinopathy of Prematurity: Natural History Data From the CRYO-ROP and LIGHT-ROP.

# **ROP Tracking List**

NOTE: To use as Excel document, click on the list, choose “Worksheet Object” and then “Open.”



# **ICROP. Synopsis of International Classification of Retinopathy of Prematurity Revisited (ICROP 2005)[[2]](#footnote-2)**

* UNIFYING PRINCIPLES UNDERLYING CLASSIFICATION
	+ The more posterior the disease and the greater the amount of avascular retinal tissue, the more serious the disease
* REVISIONS incorporated into the 2005 recommendations
	+ Concept of a more virulent retinopathy usually observed in the lowest-birth-weight infants—aggressive posterior ROP (AP-ROP).
	+ Description of an intermediate level of vascular dilatation and tortuosity (pre-plus disease) between normal-appearing posterior pole vasculature and frank plus disease that has marked dilation and tortuosity of the posterior pole vessels
	+ Clarification of the extent of zone I.
* LOCATION (3 zones)
	+ Each zone is centered on the optic disc rather than the macula, in contrast to standard retinal drawings.
	+ Zone I (posterior pole or innermost zone) consists of a circle, the radius of which extends from the center of the optic disc to twice the distance from the center of the optic disc to the center of the macula.
	+ Zone II extends centrifugally from the edge of zone I to the nasal ora serrata (at the 3 o’clock position in the right eye, and the 9 o’clock position in the left eye).
	+ Zone III is the residual crescent of retina anterior to zone II.
		- By convention, zones II and III are considered to be mutually exclusive.
		- ROP should be considered to be in zone II until it can be determined with confidence that the nasal-most 2 clock hours are vascularized to the ora serrata.
* EXTENT OF DISEASE (clock hours)
	+ This is specified as hours of the clock or as 30° sectors. As the observer looks at each eye, the 3 o’clock position is to the right and nasal in the right eye, and temporal in the left eye, and the 9 o’clock position is to the left and temporal in the right eye, and nasal in the left eye.
	+ The boundaries between sectors lie on the clock hour positions; that is, the 12-o’clock sector extends from 12 o’clock to 1 o’clock.
* STAGING OF THE DISEASE: 5 stages
	+ Describes the abnormal vascular response at the junction of the vascularized and avascular retina.
		- Because more than one ROP stage may be present in the same eye, staging for the eye as a whole is determined by the most severe manifestation present. For purposes of recording the complete examination, each stage is defined and the extent of each stage by clock hours or sector is recorded.
	+ Stage 1: Demarcation Line
		- This line is a thin but definite structure that separates the avascular retina anteriorly from the vascularized retina posteriorly.
		- There is abnormal branching or arcading of vessels leading up to the demarcation line that is relatively flat, white, and lies within the plane of the retina.
		- Vascular changes can be apparent prior to the development of the demarcation line, such as dilatation rather than tapering of the peripheral retinal vessels, but these changes are insufficient for the diagnosis of ROP.
	+ Stage 2: Ridge
		- The ridge is the hallmark of stage 2 ROP. It arises in the region of the demarcation, has height and width, and extends above the plane of the retina. The ridge may change from white to pink and vessels may leave the plane of the retina posterior to the ridge to enter it.
		- Small isolated tufts of neovascular tissue lying on the surface of the retina, commonly called “popcorn,” may be seen posterior to this ridge structure. Such lesions do not constitute the fibrovascular growth that is a necessary condition for stage 3.
	+ Stage 3: Extraretinal Fibrovascular Proliferation
		- Extraretinal fibrovascular proliferation or neovascularization extends from the ridge into the vitreous. This extraretinal proliferating tissue is continuous with the posterior aspect of the ridge, causing a ragged appearance as the proliferation becomes more extensive.
		- The severity of a stage 3 lesion can be subdivided into mild, moderate, or severe depending upon the extent of extraretinal fibrovascular tissue infiltrating the vitreous.
	+ Stage 4: Partial Retinal Detachment
		- Stage 4 is divided into extrafoveal (stage 4A) and foveal (stage 4B) partial retinal detachments. Stage 4 retinal detachments are generally concave and most are circumferentially oriented.
		- The extent of retinal detachment depends upon the number of clock hours of fibrovascular traction and their degree of contraction.
		- Typically, retinal detachments begin at the point of fibrovascular attachment to the vascularized retina. In progressive cases, the fibrous tissue continues to contract and the tractional retinal detachment increases in height, extending both anteriorly and posteriorly.
		- Radial detachments and more complex configurations are less common.
	+ Stage 5: Total Retinal Detachment
		- Retinal detachments are generally tractional and may occasionally be exudative.
		- They are usually funnel shaped. The configuration of the funnel itself permits a subdivision of this stage. The funnel is divided into anterior and posterior parts.
			* When open both anteriorly and posteriorly, the detachment generally has a concave configuration and extends to the optic disc.
			* The funnel can be narrow in both its anterior and posterior aspects with the detached retina located just behind the lens.
			* The funnel can be open anteriorly but narrowed posteriorly (less common).
			* The funnel can be narrow anteriorly and open posteriorly (least common).
* PLUS DISEASE
	+ The above stages focus on the changes at the leading edge of the abnormally developing retinal vasculature.
	+ Additional signs indicating the severity of active ROP have been referred to as “plus” disease. These include:
		- Increased venous dilatation and arteriolar tortuosity of the posterior retinal vessels
		- Iris vascular engorgement
		- Poor pupillary dilatation (rigid pupil)
		- Vitreous haze.
	+ The definition of plus disease has been refined to define the minimum amount of vascular dilatation and tortuosity using “standard” photographs and the number of quadrants involved.
	+ A + symbol is added to the ROP stage number to designate the presence of plus disease.
		- Stage 2 ROP combined with posterior vascular dilatation and tortuosity would be written “stage 2+ ROP.”
* PRE-PLUS DISEASE
	+ ROP activity indicated by abnormal dilatation and tortuosity of the posterior pole vessels. Plus disease is the severe form of this vascular abnormality.
	+ Pre-plus disease is defined as vascular abnormalities of the posterior pole that are insufficient for the diagnosis of plus disease but that demonstrate more arterial tortuosity and more venous dilatation than normal.
	+ Over time, the vessel abnormalities of pre-plus may progress to frank plus disease as the vessels dilate and become more tortuous.
	+ Note pre-plus after the stage: “stage 2 with pre-plus disease.”
* AGGRESSIVE POSTERIOR ROP
	+ This is an uncommon, rapidly progressing form designated AP-ROP. **If untreated, it usually progresses to stage 5 ROP.**
	+ It is characterized by:
		- Posterior location
		- Prominence of plus disease
		- Ill-defined nature of the retinopathy.
	+ Most common in zone I, but may occur in posterior zone II
	+ Development and distinguishing features
		- Early on, posterior pole vessels show increased dilation and tortuosity in all 4 quadrants that is out of proportion to the peripheral retinopathy
		- The vascular changes progress rapidly
		- Shunting occurs from vessel to vessel within the retina and not solely at the junction between vascular and avascular retina
		- Often difficult to distinguish between arterioles and venules because both have significant dilation and tortuosity
		- May be hemorrhages between vascularized and avascular retina
		- Does not progress through the classic stages 1 to 3
		- May appear as only a flat network of neovascularization at the deceptively featureless junction between vascularized and nonvascularized retina and may be easily overlooked
		- Typically extends circumferentially and is often accompanied by a circumferential vessel
		- Performing indirect ophthalmoscopy with a 20-D condensing lens instead of a 25- or 28-D lens may help to distinguish the deceptively featureless neovascularization
	+ Previously referred to as “type II ROP” and “Rush disease.” Aggressive, posterior ROP more accurate.Diagnosis can be made on a single visit, does not require evaluation over time.
* REGRESSION OF ROP
	+ Most ROP regresses spontaneously by a process of involution or evolution from a vascoproliferative phase to a fibrotic phase
	+ One of the first signs of stabilization of the acute phase of ROP is the failure of the retinopathy to progress to the next stage.
	+ Morphological signs of regression
		- Occurs largely at the junction of vascular and avascular retina as retinal vascularization advances peripherally
		- On serial examinations, the anteroposterior location of retinopathy may change from zone I to zone II or from zone II to zone III.
		- The ridge may change in color from salmon pink to white.
	+ Involutional sequelae of ROP
		- Peripheral changes
			* Vascular
				+ Failure of peripheral, retinal vascularization
				+ Abnormal, nondichotomous branching of the retinal vessels
				+ Vascular arcades with circumferential interconnection
				+ Telangiectatic vessels
			* Retinal
				+ Pigmentary changes
				+ Vitreoretinal interface changes
				+ Thin retina
				+ Peripheral folds
				+ Vitreous membranes with or without attachment to retina
				+ Lattice-like degeneration
				+ Retinal breaks
				+ Traction-rhegmatogenous retinal detachment
		- Posterior changes
			* Vascular
				+ Vascular tortuosity
				+ Straightening of blood vessels in temporal arcade
				+ Decrease in angle of insertion of major temporal arcade
			* Retinal
				+ Pigmentary changes
				+ Distortion and ectopia of macula
				+ Stretching and folding of retina in macular region leading to periphery
				+ Vitreoretinal interface changes
				+ Vitreous membranes
				+ Dragging of retina over optic disc
				+ Traction-rhegmatogenous retinal detachment
			* The more severe the acute phase of the retinopathy, the more likely involutional changes will be severe as the disease enters what was formerly called the “cicatricial phase.”
1. “Screening Examination of Premature Infants for Retinopathy of Prematurity.” Policy Statement (PS) issued by the American Academy of Pediatrics (AAP) Section on Ophthalmology, the American Association of Pediatric Ophthalmology and Strabismus (AAPOS), and the American Academy of Ophthalmology (AAO). Originally issued in 1997 and updated in 2001, 2005, and 2006; current version published in *Pediatrics* (Volume 131, Number 1, 2013, at <http://pediatrics.aappublications.org/content/131/1/189>. This document refers to recommendations based upon the numbers assigned to them in the PS. [↑](#footnote-ref-1)
2. The International Classification of Retinopathy of Prematurity Revisited. International Committee for the Classification of Retinopathy of Prematurity. *Arch Ophthalmol* 2005. 123: 991-999. Available at [ICROP](https://jamanetwork.com/journals/jamaophthalmology/fullarticle/417157). [↑](#footnote-ref-2)