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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 5. Transfer for non-ROP care**

**Transfer for non-ROP care principles**

1. The hospital may only transfer an infant who needs ROP care if it first:
   1. Obtains the agreement of the transferring, hospital-based ophthalmologist **AND**
   2. Schedules ophthalmic care at the receiving hospital with an ophthalmologist who agrees to screen for ROP **AND**
   3. Verifies that the receiving hospital has designated someone who is familiar with and understands the ROP Screening Policy Statement to track the ROP care until the infant meets the end-of-acute screening criteria [[Table 5. When to stop](#_Table_5._)] **AND**
   4. Confirms that the hospital will schedule the first outpatient ROP appointment if the infant is discharged before ROP screening is complete **AND**
   5. Sends the hospital appropriate records and current contact information for the parent.

**Transfer process**

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

1. The neonatologist notifies the ophthalmologist and hospital ROP coordinator (H-ROPC) that a transfer for non-ROP care is planned, and clarifies whether the infant will be transferred back after the non-ROP care.
2. The ophthalmologist determines if the infant needs another ROP exam or additional treatment prior to transfer and writes a final ophthalmic consult note that summarizes the infant’s current ROP status and screening/treatment recommendations (a new note may not be needed if the ophthalmologist has evaluated or treated the infant very recently).
3. The ophthalmologist tells the H-ROPC and office ROPC:
   1. The interval and approximate date of the next ROP exam if screening is not complete (e.g., follow-up exam in 2 weeks around 9/25/18) [[Table 3. Follow-up exams](#_Table_3.__1)].
   2. If ROP screening is complete [[Table 5. When to stop](#_Table_5._)].
4. The ophthalmologist instructs the ROPCs to update the Hospital [ROP Tracking List](#_Tracking_list) to show that the infant was transferred for non-ROP care.
5. The ophthalmologist completes and signs the [Transfer for other care letter](#_Transfer_for_other) or [Spanish transfer for other care letter](#_Carta_de_remisión), and writes an order for the H-ROPC or NICU nurse to:
   1. Review the letter with the parent, and obtain the parent’s signature.
   2. Give a copy of the signed document to the parent.
   3. Place a copy of the signed document in the infant’s medical record.
6. The neonatologist explicitly addresses eye care in the neonatology discharge summary based upon the most recent ophthalmology note:
   1. States the interval and approximate date of the next exam (e.g., ROP exam needed in two weeks around 9/25/18) if ROP screening is not complete **OR**
   2. Directs the pediatrician to refer the infant to an ophthalmologist to screen for conditions common in premature infants, such as amblyopia, strabismus, etc.
7. The H-ROPC coordinates the transfer:
   1. Confirms that the ophthalmologist has been notified of the transfer and has agreed to it.
   2. Contacts the Admitting Nurse at the receiving hospital and:
      1. Confirms that an ophthalmologist has agreed to take over the ROP care if it is still needed,
      2. Verifies that the receiving hospital has designated someone who is familiar with and understands the ROP Screening Policy Statement to track the ROP care until the infant meets the end-of-acute screening criteria [[Table 5. When to stop](#_Table_5._)]
      3. Confirms that the hospital will schedule the first outpatient ROP appointment if the infant is discharged before ROP screening is complete
      4. Indicates the interval and approximate date of the first ROP exam at the receiving hospital,
      5. Schedules the initial ROP exam with an ophthalmologist, and
      6. Sends all pertinent medical records and current contact information for the parents.
   3. Informs the parent of the name of the ophthalmologist at the receiving hospital.
8. The H-ROPC coordinates the transfer back to the original hospital if the infant will return there:
   1. Speaks to the nurse at the other hospital to obtain the ophthalmic records, and learn the infant’s ROP status, and the interval and approximate date of the next ROP exam.
9. The H-ROPC informs the O-ROPC and the ophthalmologist who will take over when the infant is coming back, provides the other hospital’s ROP records, and schedules the next exam. The ROPCs resume tracking.

# [**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 an Excel document, click on the list, choose “Worksheet Object” and then “Open.”



# **Transfer for other care letter**

Ophthalmologist: Place on your letterhead. Revise the letter as needed if the infant will not return to the original hospital after treatment.

Dear \_\_\_\_\_\_\_\_\_

I am an ophthalmologist (eye physician and surgeon). Your baby’s doctor asked me to examine the baby’s eyes. This letter will explain why I needed to do the exam, and why the baby will need more eye exams.

**Your baby may have a condition of the retina (the back of the eye) called ROP (retinopathy of prematurity).** After a premature birth, the blood vessels at the back of the eye may stop growing. The baby’s body responds by making a chemical called VEGF (vascular endothelial growth factor). This chemical makes new blood vessels start growing.

But these are not normal blood vessels. These abnormal blood vessels can bleed. They can also pull (detach) the retina away from its normal position. This is called an RD (retinal detachment), and it can cause blindness. ROP needs to be treated with 72 hours if it reaches a certain stage. Your baby could go blind without treatment.

**The next few months are very important.** We need your help to keep your baby from going blind. An ophthalmologist will need to examine the baby’s eyes many times. The ophthalmologist is checking for abnormal blood vessels. The exams must continue until the blood vessels heal. An ophthalmologist at the next hospital will examine the baby’s eyes there. I will examine the baby’s eyes again at this hospital if your baby comes back here. Some exams may be needed after you take the baby home.

When your baby is allowed to go home, the hospital will make an ROP appointment with an ophthalmologist. You must bring the baby in to the office or clinic for every appointment. The ophthalmologist will contact you if you missan appointment. If the ophthalmologist cannot reach you, the ophthalmologist may need to contact Child Protective Services to help bring the baby in for an eye exam.

**Here is what I found today when I examined your baby**

* \_\_\_\_Your baby’s blood vessels are abnormal. The baby may need treatment soon. An ophthalmologist at the next hospital will examine the baby each week to see if treatment is needed. The next ROP exam needs to take place in \_\_\_\_\_ weeks around \_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_ (date).
* \_\_\_\_Your baby’s blood vessels are abnormal. But the baby does not need treatment right now. An ophthalmologist at the next hospital will examine the baby again to see if treatment is needed. The next ROP exam is on \_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_ (date) in \_\_\_\_\_ weeks.
* \_\_\_\_\_Your baby’s blood vessels are almost normal. The baby will not need treatment for ROP. But the baby needs a different type of eye exam. This exam will include a check for crossed eyes, lazy eye, or nearsightedness. Your baby needs to be checked on about \_\_\_\_\_\_\_\_\_\_\_ (date). Ask the baby’s doctor (pediatrician) for a referral to an ophthalmologist. Then call the ophthalmologist and make the appointment.

\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_ Name of ophthalmologist \_\_\_\_\_ Date

# **Carta de remisión a otro centro de atención**

Para el oftalmólogo: Le agradecemos copiar esta carta en su papel membreteado. Lea la carta y haga los cambios necesarios si el (la) bebé no tiene que regresar al hospital original después del tratamiento.

Apreciado(a) \_\_\_\_\_\_\_\_\_

Soy un oftalmólogo (médico y cirujano de ojos). El doctor que ve a su bebé me pidió que le examinara los ojos. Esta carta explicará la razón por la cual necesito realizar el examen y por qué el (la) bebé va a requerir otros exámenes oculares adicionales.

**Su bebé puede tener una afección de la retina (la parte posterior del ojo) que se conoce como ROP (retinopatía de la prematurez).** Después de un nacimiento prematuro, los vasos sanguíneos de la parte posterior del ojo pueden dejar de crecer. El cuerpo del bebé responde produciendo una sustancia química que se conoce como VEGF (factor de crecimiento vascular endotelial). Esta sustancia química hace crecer nuevos vasos sanguíneos.

Pero estos vasos sanguíneos no son normales. Los vasos sanguíneos anormales pueden sangrar. Además pueden halar (desprender) la retina de su posición normal. Esto es lo que se conoce como DR (desprendimiento de retina) y puede terminar en ceguera. La ROP tiene que tratarse en el término de 72 horas si llega a un determinado nivel. Su bebé podría quedar ciego(a) a menos que reciba tratamiento.

**Los próximos meses son muy importantes.** Tenemos que ayudar a impedir que su bebé pierda la vista. Un oftalmólogo tendrá que examinarle los ojos muchas veces. Lo que el oftalmólogo verifica es si hay desarrollo de vasos anormales. El examen debe seguir haciéndose cada cierto tiempo hasta que los vasos sanguíneos estén sanos. Un oftalmólogo en otro hospital examinará los ojos de su bebé. Yo lo (la)examinaré de nuevo en este hospital si el otro médico considera que debe volver aquí (en ese caso, deberá traer aquí de nuevo a su bebé. Es posible que se requieran más exámenes después de que lleve a su bebé a casa.

Una vez que el (la) bebé salga del hospital para la casa, el hospital programará una cita de ROP con un oftalmólogo. Debe traer al bebé al consultorio o a la clínica para cada cita. El oftalmólogo se comunicará con usted en caso de que no cumpla una cita. Si el oftalmólogo no puede comunicarse con usted, es posible que tenga que contactar a los Servicios de Protección del Menor para ayudar a que podamos examinar los ojos del (de la) bebé.

**Lo siguiente es lo que he encontrado al examinar hoy a su bebé**

* \_\_\_\_ Los vasos sanguíneos de su bebé son anormales. Es posible que el (la) bebé requiera tratamiento a la mayor brevedad. Es necesario que lo (la) examine un oftalmólogo en el hospital semanalmente para ver si requiere tratamiento. El siguiente examen de ROP debe realizarse en \_\_\_\_\_ semanas aproximadamente \_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_ (fecha).
* \_\_\_\_ Los vasos sanguíneos de su bebé son anormales. Pero el (la) bebé no requiere tratamiento de inmediato. En el siguiente hospital lo (a) examinará otra vez un oftalmólogo para ver si se requiere tratamiento. El siguiente examen de ROP se hará el \_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_ (fecha) en \_\_\_\_\_ semanas.
* \_\_\_\_\_Los vasos sanguíneos de su bebé están casi normales. el (la) bebé no requerirá tratamiento para la ROP. Sin embargo, requiere un tipo diferente de examen ocular. Este examen incluirá un estudio para determinar si es bizco(a), si tiene un ojo perezoso o si es miope. Su bebé deberá ser examinado(a) en una cita de control aproximadamente el \_\_\_\_\_\_\_\_\_\_\_ (fecha). Pídale a la médico (pediatra) de su bebé que lo (la) remita a un oftalmólogo. Después llame al consultorio del oftalmólogo para pedir una cita.

\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_\_ Nombre del oftalmólogo \_\_\_\_\_ fecha

# **ICROP. Synopsis of International Classification of Retinopathy of Prematurity Revisited (ICROP 2005)[[1]](#footnote-1)**

* 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. 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 <https://jamanetwork.com/journals/jamaophthalmology/fullarticle/417157>; accessed 6/25/18. [↑](#footnote-ref-1)