Retinopathy of Prematurity (ROP)

What is Retinopathy of Prematurity?
Retinopathy of Prematurity (ROP) is a retinal condition that affects premature infants. The retina begins to develop at about 16 weeks after conception and continues to fully develop prior to a full term birth (typically 37-42 weeks). When an infant is born prematurely, the development of blood vessels within the eye is interrupted, causing undeveloped areas of avascular tissue (see picture above). The amount of avascular tissue present, and consequently the extent of ROP, is dependent on how premature the infant is. Typically, the younger an infant is born and the less it weighs, the more avascular retina is present.

Even after birth, the retina attempts to continue developing. In response to low oxygen levels within the avascular tissue, a chemical signal called Vascular Endothelial Growth Factor (VEGF) is released in an effort to extend the existing blood vessels into this area. However, since there was an interruption in normal development, the new blood vessels that grow in response to the VEGF are abnormal and grow without direction, which can form a visible ridge on the retina (see picture above, white arrow). In some cases, these abnormal vessels at the ridge can leak blood into the vitreous cavity in front of the retina. This blood contracts and pulls on the retina causing a tractional retinal detachment and permanent blindness if left untreated.
What are the risk factors?

Retinopathy of Prematurity (ROP) only occurs in premature babies. The two most significant risk factors for the development and severity of ROP is gestational age (number of weeks after conception) and birth weight. In most hospitals, it is policy that all babies with a gestational age of 32 weeks or less and/or a birth weight of 1,200 grams or less be screened for ROP. Though only a small percentage of infants with a gestational age of 32 weeks and/or a birth weight of 1,200 grams have ROP, the percentage of infants with ROP significantly increases with lower gestational ages and/or birth weights.

Another risk factor for the development of ROP is an unstable clinical course, even if an infant has a gestational age above 32 weeks and/or a birth weight above 1,200 grams. An unstable clinical course includes the presence of severe respiratory (bronchopulmonary dysplasia) or heart (patent ductus arteriosis) disorders, the necessity of being on a respirator, and/or a bleed into the brain (intraventricular hemorrhage).

How can the doctor determine the extent of Retinopathy of Prematurity?

Screening for ROP begins in the NICU, and once an infant is identified as being at risk, consultation from a qualified ophthalmologist is sought. In order to perform the eye exam, the infant’s eyes must be fully dilated, which typically takes around an hour after the drops are placed in the eyes. The dilation drops used are harmless and the effects of the drops wear off after about 2-4 hours (depending on eye color). Once the infant’s eyes are fully dilated, the doctor will use an instrument called an indirect ophthalmoscope along with a high powered lens to examine the retina. The exam itself only takes around 10 minutes and the infant’s eyes will be numbed to ensure that the infant will be comfortable through-out the procedure. If leaking blood vessels are found, or if there is a large number of abnormal blood vessels, the doctor may order Fluorescein Angiography.

How is Retinopathy of Prematurity classified?

To standardize communication about ROP between health care professionals, an international committee created a specific method for diagnosing and categorizing the extent of ROP present. This classification can be broken down into three main categories: zone, stage, and presence of plus disease.

- **Zone** refers to the location of where the avascular tissue starts in the retina in relation to the optic nerve. ROP within zone 1 (area closer to the optic nerve) tends to create the most complications, whereas ROP in zone 3 (area far from the optic nerve) rarely creates complications.
- **Stage** refers to the clinical appearance of the junction between the vascular and avascular tissue. There are six stages.
  - **Stage 0** indicates that avascular tissue is present, but there is no definite line separating it from the vascularized tissue.
  - **Stage 1** indicates that there is a definite line separating the avascular and vascularized tissue.
  - **Stage 2** indicates that a raised area (ridge) is present between the avascular and vascularized tissue.
  - **Stage 3** indicates that there is bleeding into the retina and/or vitreous at the ridge.
  - **Stage 4 & Stage 5** indicate the presence of a retinal detachment of varying severities.
- **Plus disease** indicates the presence of dilated, corkscrew-shaped blood vessels within the eye. This occurs as a result of excessive amounts of Vascular Endothelial Growth Factor in the eye and is evidence of more severe ROP.

What treatments are available?

- **Pan Retinal Photocoagulation (PRP)** is a laser treatment that destroys the areas of avascular tissue in an attempt to prevent it from releasing VEGF. PRP is the first-line treatment for ROP.
- **Intravitreal injection** of Avastin into the eye is performed if the ROP does not respond to PRP treatment. Avastin is an anti-VEGF medication that prevents new blood vessel growth.
- **Pars Plana Vitrectomy** is performed if a vitreous hemorrhage or retinal detachment is present or if the ROP does not respond to the above treatments.