

Our Patient is a Premature Baby: Is an Eye Check Required? (Retinopathy of Prematurity)

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What is Retinopathy of Prematurity?

Retinopathy of prematurity (ROP) is a developmental disorder that occurs in the incompletely vascularised retina of premature infants and is an important cause of blindness in children in both the developed and the developing countries.

Why should we worry?

Incidence of this condition is rising rapidly in developing countries with improvements in neonatal care and increasing survival of very low birth infants.

Out of 26 million annual live births in India, approximately 2 million are at risk of developing ROP.

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In India the incidence of ROP is between 38 and 51.9% in low-birth-weight infants.¹

Lack of awareness among ophthalmologists and neonatologists is leading to a huge increase in ROP related blindness, which poses a huge socio-economic burden on the community.

History

ROP was first described by Terry in the 1940s.

1942: Terry identified the disease & coined the term 'Retrolental Fibroplasia'.

1951: Campbell suggested that the toxic effects of oxygen in the neonates caused the disease.

1952: Heath coined the term Retinopathy of Prematurity, which became the more accepted nomenclature.

1953: Studies proved that high concentrations of oxygen were causative.^{2,3}

Some of the Obstacles That should be Addressed Include

- Lack of awareness of ROP and its outcome among parents and primary care physicians
- Lack of dedicated ambulance service for sick new-borns, (Mobile van units for ROP screening)
- Non-adherence to agreed patient pathways, and shortage of nurses and ROP trained ophthalmologists

WHO has recognised that prematurity is one of the major causes of neonatal mortality (Report - WHO: Born Too Soon 2012) with India having largest number of premature babies (3, 59,100 premature babies annually) in the world.

ROP in India is severe in its form and is seen in higher birth weight groups too.^{4,5}

Screening for ROP should be performed in all preterm neonates who are

- Born < 34 weeks gestation and/or < 1750 grams birth weight;
- 34-36/7 weeks gestational age
- 1750-2000 grams birth weight
- Infants born < 28 weeks or < 1200 grams birth weight should be screened early, by 2-3 weeks of age, to enable early identification of AP-ROP

The first retinal examination should be performed not later than 3 weeks of age or 30 days of life in infants born < 28 weeks of gestational age.⁶

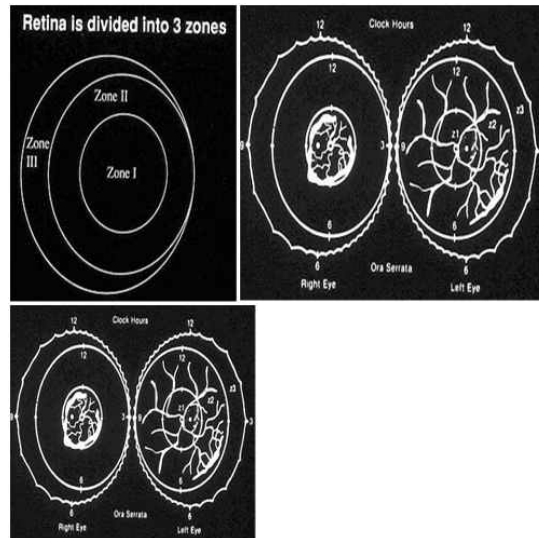
Risk Factors For Rop

- Prematurity and Low birth weight
- High oxygen levels
- Septicaemia
- Acidosis
- Hyperglycaemia
- Multiple gestation
- Anaemia
- Respiratory distress syndrome
- Apnoeic spells
- Intraventricular haemorrhage.^{1,2,3,4,6,8}

Classification

A committee for ROP classification was formed in 1984, which proposed an

international classification of ROP (ICROP) by dividing the retina into three zones, extending from posterior to anterior retina and the extent of ROP in clock-hours of involvement.



Zones

Three concentric zones, centred on the retina define the antero-posterior location of retinopathy.

Zone I: With optic disc as the centre, and twice the distance from the disc to fovea, the circle formed is zone I.

Zone II: It starts from edge of zone I and extends till the ora serrata nasally, with a corresponding area temporally.

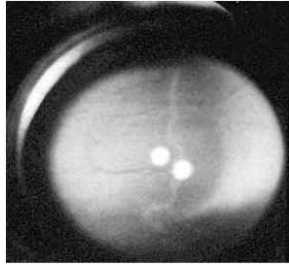
Zone III: Zone III is the remaining crescent of retina temporally.⁹

Extent of retinopathy

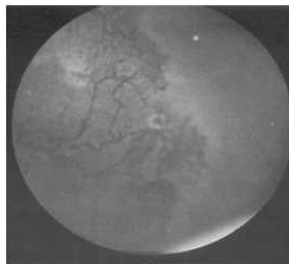
The extent of the ROP is documented by the number of clock hours involved.

Stages of ROP

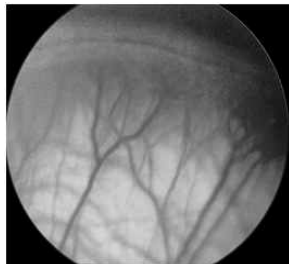
Stage 1 - A **demarcation line** is seen between the vascular and avascular retina. It is a thin structure that lies in the plane of the retina



Stage 2 - The demarcation line grows to occupy a volume and has a height and width to form a ridge above the plane of retina. Small tufts of new vessels called as "popcorn" vessels may be seen posterior to the ridge.

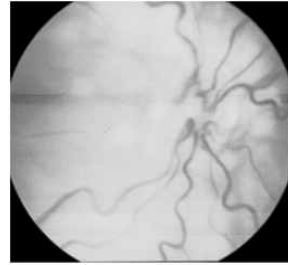


Stage 3 - In this stage extraretinal fibrovascular tissue is seen arising from the ridge into the vitreous.

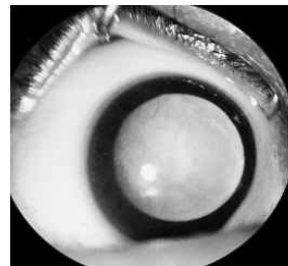


Stage 4 - A partial detachment of the retina is seen which may be exudative or tractional. It is sub divided into the following:

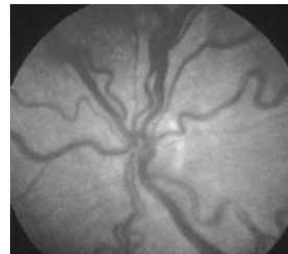
- (1) Partial retinal detachment not involving the fovea (**stage 4A**)
- (2) Partial retinal detachment involving the fovea (**stage 4B**)



Stage 5 - A total retinal detachment is seen as child usually presents with leucocoria (white pupillary reflex.)



Plus disease: It is an indicator of severity of the disease and is defined as venous dilation and arterial tortuosity of posterior pole vessels.

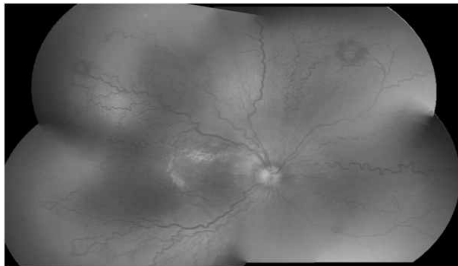


Pre-plus disease: It is defined as posterior pole vascular dilation and tortuosity which is more than normal but less than plus disease.

Aggressive posterior ROP: (APROP) This refers to rapidly progressive, form of ROP referred to as "rush disease".

It is characterised by a posterior location, severe plus disease, and flat intraretinal neovascularisation. It can progress very fast to stage 5 ROP, if not

intervened early.⁹



Examination Technique

- The examination technique involves the dilatation of pupil and indirect ophthalmoscopy preferably with a 20 D/28D lens.
- Perform pupillary dilatation 45 min prior to commencement of the screening.
- Dilating drops used are a mixture of tropicamide (0.4%) and phenylephrine (2.5%) drops to be applied two to three times about 10-15 min apart.
- The nurse should be instructed to wipe any excess drops from the eye lid to prevent systemic absorption.
- If the pupil is resistant to dilatation, it may indicate presence of persistent iris vessels (tunica vasculosa lentis) and must be confirmed by the ophthalmologist before applying more drops.

Treatment for ROP

Threshold ROP: The cryotherapy for retinopathy of prematurity (CRYO-ROP) study stated that treatment should be imparted to eyes with threshold disease, defined as stage 3 ROP in zone I or II, having five contiguous or eight discontinuous clock hours with plus disease. This was the previous "cut off" for treatment.

Pre-threshold ROP: The early treatment for retinopathy of prematurity (ETROP) study redefined these guidelines. They defined the actively treatable and observational types of pre-threshold ROP as "type 1" (high-risk prethreshold ROP) and "type 2" ROP respectively.

"Type 1 ROP" is defined as: (1) Any stage of ROP in zone I with plus disease or (2) Stage 3 in zone I without plus; or (3) Stages 2 or 3 in zone II with plus disease. These are the modified guidelines for treatment.

"Type 2 ROP" is defined as stages 1 or 2 in zone I without plus, or stage 3 in zone II without plus. These can be observed and watched at one week or less follow-up.

Cases having stages 1 or 2 in zone II require two weekly follow up, while stages 1 or 2 in zone III require three weekly follow-ups.^{10,11,12}

Problems in ROP Screening

- All screening programmes are time-consuming and labour-intensive, uncomfortable to the infants, cause anxiety to the parents and sometimes lead to an extended stay at nurseries.
- All these factors have to be weighed against missing a child with treatable ROP.
- Medicolegal implications are more pertaining to diagnosis and management.
- ROP screening is more of a team effort and requires efforts of Neonatologist and Ophthalmologist with nursing staff.

Medico legal implications

- Screening for ROP needs to be initiated timely after birth to prevent blindness.

- It is the responsibility of the caring paediatrician to initiate screening by referring to an ophthalmologist and it is the responsibility of the ophthalmologist to do correct screening and treatment.

This has immense medico legal implications because if a child goes blind due to missed or late screening then the paediatrician and the ophthalmologist are at a very high risk of getting into a law suit.

How can we be a part of the cause?

- National Health Mission and the Rashtriya Bal Swasthya Karyakram (RBSK) has set up a task force of all the experts to set up the programme for the whole country.
- The Indian Retinopathy of Prematurity (IROP) Society was initiated in July 2016 by some of the leading experts in the field to bring together ROP specialists from India to share best case practices, promote uniform guidelines, enhance screening efforts into unreached areas through training and technology, create a medico-legal framework for ethical and clinical excellence in ROP management. 7
- Tele screening techniques are now employed to reduce this burden

Comorbidities

Association with co morbid conditions affecting the lungs, intestines, brain and association with several pervasive developmental disorders need special attention to make the kid's life comfortable.

Take Home Message

1. ROP is a preventable disease
2. If detected early it is completely

salvagable

3. Even in the fulminant apro cases early rehabilitation is a must

Follow up examinations should be based on the retinal findings and should continue until complete vascularisation or regressing ROP is documented or until treated based on the ETROP guidelines. LASER photocoagulation delivered by the indirect ophthalmoscopic device/ Intravitreal anti-VEGF are the options for ROP treatment.



4. If referred late surgery is the answer with decreasing prognosis as the stage of ROP goes higher

The responsibility of recognition of infants for screening lies with the paediatrician/neonatologist. Communication with the parents regarding timely screening, seriousness of the issue is extremely important. The refraction has to be done at the 6th month as a major section of these children have myopia which needs correction to aid their

overall global development



5. The main challenge is to reach this baby within 30 days.
6. The child is not born with the ROP and can be treated with timely intervention.
7. ROP occurs 2-3 weeks after birth so there is a window of opportunity for treatment.

Message for General Practitioners

Being the closest to any given family unit, he is equipped to advise any family on the necessity to get a timely examination done in case of a preterm birth in the household and to apprise them on the pitfalls of neglecting these examinations and follow-up.

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Antidepressant utilisation and incidence of weight gain during 10 years' follow-up

The risk of weight gain remained increased during at least six years of follow-up. In the second year of treatment the number of participants treated with antidepressants for one year for one additional episode of 5% or more weight gain was 27 (95% confidence interval 25 to 29).

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