# RETINOPATHY OF PREMATURITY







# PRETERM BABIES TOO, HAVE THE RIGHT TO SIGHT

In the context of preventing blindness in newborn babies, Retinopathy of Prematurity (ROP) has emerged as a severe challenge particularly in developing and middle-income group countries in Latin America, Eastern Europe, South East Asia, China and India.

ROP is a potentially avoidable cause of irreversible and usually total blindness in preterm babies. This disease has lifelong implications for afflicted children and their families.

India continues to lead with the largest number of visually impaired and blind children, globally.

Many countries like India are expanding neonatal care but lack

sufficient knowledge, effective screening guidelines and bedside programs for ROP.

More than 60% of visual impairments in babies are preventable or curable with timely detection, prompt and appropriate preventive and curative management.

The World Health Organization (WHO) has highlighted ROP as a major target disease in its prevention of blindness program, "VISION 2020: Right to Sight" to combat needless blindness globally by the year 2020.

The program targets all babies at risk for ROP, for screening eye examinations and access to treatment for severe ROP.



# RETINOPATHY OF PREMATURITY

Retinopathy of Prematurity (ROP) is a dynamic, time-bound disease that is not present at birth.

The condition afflicts the eyes of preterm babies that have typically received hospital based neonatal care (with or without oxygen therapy) that helps to save their life, but severely affects their eye development.

The condition is characterized by development of abnormal blood vessels in the retina of the eye, resulting in scarring and retinal detachment.

ROP can be mild and may resolve spontaneously, but in serious cases, may progress rapidly and lead to blindness.

ROP typically starts only 2-3 weeks after birth, providing a window period for screening and initiating treatment at the right time, while the baby is still under neonatal care at the hospital.



### **40 CRITICAL WEEKS**

The last twelve weeks of a normal term gestation are crucial in the development of fetal eyes.

The fetal retina (tissue that lines the back of the eye) slowly matures in the mother's womb by 40 weeks, which is by the expected date of delivery.

From 16 weeks to birth, retinal blood vessels grow out from the optic nerve to reach the peripheral retina.

In preterm babies, the normal growth of blood vessels stops. The area without adequate blood

supply emits a chemical trigger to stimulate growth of abnormal vessels.

These abnormal blood vessels are fragile and can lead to formation of a ring of scar tissue that is attached to both the retina and the vitreous gel that fills the center of the eyes.

As the scar tissue contracts, it may pull the retina out of position, creating a retinal detachment.

Retinal detachment is the prime cause of visual impairment and blindness in ROP.



# TEES DIN ROSHNI KE<sup>\*</sup>!

Chronic hypoxia (lack of oxygen), intrauterine growth retardation and prenatal and postnatal conditions are the most common triggers of ROP.

Babies born under 34 weeks gestation and weighing less than 2000 grams are particularly susceptible to ROP and must be screened within 20-30 days from birth.

High levels of supplemental oxygen and high carbon dioxide levels are also known to aggravate ROP. During neonatal incubation, preterm babies are to be provided

<sup>\*</sup>Thirty days to vision!

with blended oxygen, strictly controlled and monitored using pulse oxymeters.

Other risk factors that are associated with the condition include anemia, bradycardia (low heartrate), blood transfusions and intraventicular hemorrhage (bleeding into the brain).

Additionally, prenatal maternal factors compound the probability of an ROP occurrence. These include infertility treatments, twins and triplets, heavy smoking, a n e m i a, d i a b e t e s a n d preeclampsia.



## BORN TOO SOON

Early stages of ROP, where treatment is effective, have no symptoms and the eye looks normal from the outside.

Hence, routine retinal screening within 3-4 weeks of birth is the only way to detect vision-threatening stages of ROP.

- Preterm babies born at 23-27 weeks should be examined within three weeks of birth.
- Preterm babies born at or beyond 28 weeks should be examined by the fourth week, by Day-30 of life.
- Preterm babies should be examined prior to discharge from the hospital if they are likely to miss a follow-up examination.

During this period, most preterm babies are in critical care and often incubated in Neonatal Intensive Care Units (NICU) and Special Newborn Care Units (SNCU).

An eye specialist must be appointed at the neonatal care hospital, to examine the retina of newborns. Caregivers need to be geared up to conduct ROP screening and treatment in such adverse situations.

For positive outcomes, treatment with laser photocoagulation must be carried out within 72 hours of detection of the condition.

Follow-up screening as recommended after initial examination must be strictly followed to avoid irreversible vision loss.

All preterm children run a higher risk in developing other eye and visionrelated complications later in their lives.

Even preterm children without ROP condition are likely to develop squint, lazy eyes (amblyopia) and significant refractive problems that require prescription eyeglasses.

Thus, bi-annual eye examinations are recommended for all preterm babies born under 34 weeks or weighing less than 2000 grams.

The most effective prevention of ROP is the prevention of premature birth that is currently not possible. In fact these instances are on the rise partially due to assisted fertilization techniques that often result in premature and multiple births.

Preventing other complications of prematurity (such as neonatal respiratory distress syndrome) may also help prevent ROP. Antenatal steroids administered to high-risk mothers are one of the options available.

# NATIONAL ROP TASK FORCE (2014 - TILL DATE) Ministry of Health and Family Welfare

- Chair: 1) Smt Vandana Gurnani, Joint Secretary (RMNCH+A), Ministry of Health and Family Welfare
  - 2) **Dr R Azad**, Ophthalmology expert, Ex Director, Dr R.P. Centre for Ophthalmic Sciences, All India Institute for Medical Sciences (AIIMS), New Delhi

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### **Expert Working Group**

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- Dr Srinivas Murki, Fernandez Hospital, Hyderabad
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- Dr Mangat Dogra, PGIMER, Chandigarh
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- Dr Anand Vinekar, Narayana Nethralaya, Bangalore

*Convener:* **Dr Ajay Khera,** Deputy Commissioner-in-charge (Child Health and Immunization), Ministry of Health & Family Welfare



# VISION 2020: R SPOTLIGHT ON RETINOPAT

### CHILDHOOD BLINDNESS

A group of diseases and conditions occurring in childhood or early adolescence, which, if left untreated, result in blindness or severe visual impairment that are likely to be untreatable later in life.

### THE SHOCKING REALITY

**1.4 mn** blind children across the world

3 of 4 blind children live in the poorest regions of Africa and Asia

Globally has the **largest number** of blind children

Sources: Gilbert & Foster; Correlations and estimations

### **RETINOPATHY OF PREMATURITY**

ROP afflicts the eyes of preterm babies that have typically received specialized neonatal care



Characterized by development of **abnormal blood vessels in the retina** of the eye, resulting in scarring and retinal detachment



Starts 2-3 weeks after birth, providing a window period for screening and activating treatment at the right time

### IMPACT OF CHILDHOOD BLINDNESS

**Only 20% of those blind are productive**, with a rate of 25% productivity as compared to sighted individuals.

The family burden to raise a blind child is **not only** economic, but also emotional.

Source: The Societal Burden of Blindness Secondary to Retinopathy of Prematurity in Lima, Peru. October 2012. American Journal of Opthamology

### CAUSES OF CHILDHOOD BLINDNESS

Largely determined by socio-economic development, and the availability of primary health care and eye care services



**HIGH INCOME COUNTRIES**: Lesions of the optic nerve and higher visual pathways

**MIDDLE INCOME COUNTRIES**: Retinopathy of Prematurity (ROP)

**LOW INCOME COUNTRIES**: Corneal scarring from measles, vitamin A deficiency, harmful traditional eye remedies, ophthalmia neonatorum, rubella cataract

### **ROP SURGERIES AT LVPEI**



# IGHT TO SIGHT HY OF PREMATURITY (ROP)

### ROP IS EMERGING AS THE LEADING CHILDHOOD BLINDING DISEASE

### Common vision problems in newborn babies & children

below 10 years, treated at LVPEI



# 20-40%

prevalence of ROP amongst the babies screened at LVPEI

# 9-15%

ROP severity based on arrival time and type of neonatal care

# 15%

ROP babies were from smaller towns and districts, in 2000-03

# 25-28%

ROP babies were from outside big cities, in 2012-14

Trend towards more **semi-urban and district level areas. Predicted to rise rapidly** as better neonatal care is now available in such places, that helps babies to survive.

### A SERIOUS CONCERN

# **BY 2020**

ROP will be the single largest cause of needless blindess



### **BATTLING AGAINST THE ODDS**

By creating awareness things can dramatically improve in the next 5 years



More parents will demand eye evaluation of their newborns, more health care providers (nurses/doctors) will refer all premature babies on time



More eye specialists will get trained in ROP screening and management due to increased referrals and demand



There will be **up-gradation in medical curriculum** in the area of neonatal and premature baby's eyes and vision



Incidence of ROP can be reduced through **availability and affordability** of screening and curative services

## NATIONAL ANNUAL ROP SCENARIO IN RECENT YEARS



Note:

- Data collected from individual eye hospitals across India
- Surgeries performed include Laser, Vitrectomies and Anti VEGF injections

# HOSPITALS IN INDIA GEARING UP TO HANDLE COMPLICATED ROP CASES

A team of ophthalmologists very recently were challenged to perform a difficult surgery a tiny baby, just 6 weeks old and not gaining weight, suffering from a critical, potentially blinding condition - Retinopathy of Prematurity (ROP). It could have permanently destroyed the retina of both the eyes if not operated within 3 days!

When the parents of that fragile baby came to the hospital for the first time, they were doubtful whether an Indian hospital will be equipped to handle such a delicate and challenging case. The parents were told that an emergency operation was required; risks and benefits were clearly laid out. The low weight of the baby was a challenge for the team of ophthalmologists, anesthetists, nursing staff, counsellors, and the neonatologist due to a very high risk of anesthesia-related complications, even in the best of centres. The parents finally consented to the surgery, reassured by the genuine concern of the doctors for their child's vision and life.

An emergency core team was constituted. Not only premature, the baby was anaemic and suffering from poor nutrition. Counsellors coordinated with the baby's neonatologist and the parents for all the preoperative evaluation and paperwork, without tiring this tiny life.

All preparations were made within 48 hours to make the baby as fit as possible for safe anesthesia. Blood transfusion, lungs, liver function, kidney function, nutrition and electrolyte balance, cardiac care – so much to be assessed and managed at such short notice! The baby was brought to the theatre and a highly competent neonatal anesthesiologist, played the pivotal role, supported by senior colleague and the anesthesia technician.

An accomplished retinal surgeon, operated upon both the baby's eyes using microsurgical techniques. Operating upon an eye less than 16mm in size, without causing damage to critical structures like the lens and the retina, required surgical precision, dexterity and a passion for perfection. When the baby stirred out of anesthesia and cried, the whole team and the anxious administrators, counsellors and parents outside, knew that they had all collectively as a team succeeded in saving the eye sight of the baby. Less than a month later, the healthy and happy baby spread sunshine on a return visit, as she smiled at the doctor, looking up with pretty good vision in both her bright eyes!

Hospitals in India are now successfully handling ROP cases and are also treating patients from neighboring countries.



### **FIVE STAGES**

ROP manifests itself in five stages that require varied courses of management and treatment.

#### Stage-I

There is mild abnormal blood vessel growth that requires close periodic examination, but may not need any treatment.

#### Stage-II

Blood vessel growth is moderately abnormal and in some cases may need early treatment.

Stage-I and most Stage-II do not lead to blindness. However if not monitored, they can progress to more severe stages.

#### Stage-III

Blood vessel growth is severely abnormal and the newborn requires early treatment within 72 hours as this is vision threatening.

#### Stage-IV

Blood vessel growth is severely abnormal and there is a partially detached retina. Urgent surgical treatment is recommended to diminish the chances of loss of vision.

#### Stage-V

There is a total retinal detachment and only very few eyes get minimal vision even after advanced surgical treatment.







4) Retina has detached as screening was not done within 30 days & no laser treatment was given



6) Lost vision due to no ROP Screening and very late detection





### BACK FROM THE BRINK

ROP develops 2-3 weeks after birth due to prematurity and low weight; and then worsens due to many factors.

If detected by a comprehensive retinal examination within 20-30 days from birth, ROP can be controlled and managed through a variety of treatments.

Laser Therapy or Photocoagulation is the most common type of ROP surgery in which small laser beams are used to treat the peripheral retina and stop the progression.

Cryotherapy deploys freezing temperatures to scar the peripheral retina.

Laser Therapy and Cryotherapy are only performed on babies with advanced ROP, particularly Stage-II with 'plus disease' and Stage-III disease.

Research studies are currently being conducted for newer and evolved treatments using anti-VEGF injections in the eye, as a supplement or substitute to laser therapy.

For advanced stages of ROP,

treatment options include Scleral Buckle and Vitrectomy.

Scleral Buckles are usually performed on babies in ROP Stage-IV.

This involves placing a silicone band around the eye and tightening it. This keeps the vitreous gel from pulling on the scar tissue and allows the retina to flatten back down onto the wall of the eye.

Babies who have had a Scleral Buckle need to have the band released months or even years later, since the eye continues to grow; otherwise they will become nearsighted.

Vitrectomy is performed for advanced ROP at Stages-IV and V.

This involves removing the vitreous and replacing it with a saline solution.

After the vitreous has been removed, the scar tissue on the retina can be peeled back or cut away, allowing the retina to relax and lay back down against the eye wall.



# FUTURE CONCERNS

All preterm children run a higher risk in the development of eye and vision-related complications other than ROP later in their lives.

Common afflictions include retinal detachment, myopia (near-sightedness), strabismus (crossed eyes), amblyopia (lazy eye) and glaucoma.

In most cases, these conditions can be treated or controlled.

Bi-annual eye examinations are recommended for all preterm babies born under 34 weeks or for those that weigh less than 2000 grams.

# BATTLING AGAINST THE ODDS

ROP is an increasing concern amongst young parents.

Although medical innovations in ROP treatment have decreased the probability of loss of vision, unfortunately it does not always prevent it.

If an infant does not respond to ROP treatment, the disease may progress, wherein a retinal detachment could develop.

Often, this is a 'Stage-IV' manifestation in which only a part of the retina detaches and may need urgent surgery.

In a Stage-V patient, the centre of the retina or the entire retina detaches. Central vision is threatened and surgery is often recommended to reattach the retina. However, results are often not favorable, with most children remaining permanently blind.





### EXAMINATION OF RED REFLEX IN NEWBORNS

Examination of red reflex<sup>1</sup> soon after birth and periodically thereafter in EVERY NEWBORN baby is a critical milestone in the early detection of many serious eye problems. This red reflex test however cannot detect early ROP.

Only a qualified and trained eye specialist should perform ROP screening retinal examination.

Following pupillary dilation using

eye drops, the baby's retina is examined in a dimly lit or dark room, using a special lighted instrument called an indirect ophthalmoscope.

Examination of the retina of a preterm baby will determine how far the retinal blood vessels have grown and whether or not the vessels are growing flat along the wall of the eye.

<sup>1</sup>The red reflex refers to the reddish-orange reflection of light from the retina of the eye that is observed when using an ophthalmoscope or retinoscope from approximately 30 cm distance. This can also be detected by flash photography in a dark room using the cell phone camera or a regular digital camera.

NEWBORN CARE WEEK NOVEMBER 15-21



Ministry of Health and Family Welfare Government of India





