

Programs for Detecting and Treating Retinopathy of Prematurity: Role of the Neonatal Team

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High quality perinatal-neonatal care can prevent severe Retinopathy of Prematurity (ROP) in most cases. Preterm infants who do develop retinopathy can also have good visual outcomes if screening and treatment are done timely. National Neonatology Forum published clinical practice guidelines for timely screening and treatment of ROP in neonatal care units in the country in 2010. It is also listed as one of the condition under Rashtriya Bal Swasthya Karyakram for early identification and early intervention, and is currently a focus area under the National Program for Control of Blindness. Technical and operational guidelines for screening and treatment have been released. Programs like home-based neonatal care can be utilized for ensuring timely screening and follow-up of high-risk infants. Prevention, timely diagnosis and treatment requires well-coordinated teamwork between neonatologists, ophthalmologists, nurses and obstetricians. The neonatal care team should have an evidence-based screening policy, must coordinate and facilitate screening by ophthalmologist, and provide pre and post-operative support, if treatment is required. The neonatologist also has an important responsibility of educating the healthcare workers and the families. A team approach and inter-sectoral coordination are the keys to success of a national drive to decrease the burden of preventable blindness due to ROP.

Keywords: Blindness, Neonatologist, Pediatrician, Prevention, Screening.

Retinopathy of prematurity (ROP) is a potentially blinding proliferative vitreo-retinopathy affecting the eyes of premature infants. The world is currently witnessing the 'third epidemic' of ROP [1,2] and it is emerging as a major public health concern in low- and middle-income countries, including India [1]. While the first epidemic in 1940s was attributed to increased use of oxygen, the second epidemic in 1970s was related to the increased survival of premature infants in developed countries. The current epidemic, which started in the last decade, is due to increasing number and survival of preterm births in the developing world related to better availability of obstetric and neonatal services [2]. Though the number of intensive and special care neonatal units (SCNU) in both the government and the private sector has increased dramatically in a short span of time, they often suffer from inadequate monitoring and sub-optimal quality of care [3]. The incidence of ROP in India has been reported to vary between 20-51.9% in low birthweight infants [4-7]. Apart from increasing survival of preterm infants, use of unblended and unregulated oxygen, variable quality of neonatal care, paucity of trained manpower, and lack of awareness have been

implicated for the high incidence of ROP [8].

Preterm infants are not born with ROP and there is a window period of few weeks before sight-threatening ROP develops. Apart from minimizing the incidence of severe ROP by providing quality care, the special and intensive neonatal care units have the responsibility to organize timely screening for ROP because even if ROP does occur, timely treatment can provide excellent visual outcome in most cases.

EVOLUTION OF ROP-SCREENING AND CURRENT STATUS

The first reports of ROP-screening programs were published from Chandigarh and Chennai in early 1990s [4,5]. Subsequently, ROP-screening was taken up by other institutions having NICU. Though special care neonatal units have rapidly expanded in both government and private sector, ROP screening programs have remained confined to tertiary care institutions and select few hospitals in private sector. This has been because of lack of sufficient awareness amongst neonatal care teams and non-availability of trained and interested ophthalmologists for this activity. We previously reported that 86.4% of infants presenting with stage 5

ROP had never been screened, and nearly 74% of infants were brought by parents upon noticing that the child was unable to see [9]. Pediatricians had referred none of these infants and only 25.8% were referred by an ophthalmologist [9]. On the other hand, individual efforts by few highly motivated ophthalmologists have been able to establish excellent programs in selected urban and rural areas of Hyderabad and Karnataka, respectively [10,11]. However, the scenario has started to change in the last couple of years.

National Guidelines

National Neonatology Forum (NNF) came out with evidence-based clinical practice guidelines for ROP-screening and treatment in 2010 to help and guide the neonatal and ophthalmic community in establishing consistent and quality screening, and treatment programs [12]. These guidelines pointed out that the screening cut-offs and timing for screening have to be different for our country as compared to USA and other developed countries [13]. It was recommended that all infants weighing ≤ 1750 g at birth and/or born at <34 weeks gestation should be screened for ROP. Infants with birth weight of 1750-2000 g or gestation of 34-36 weeks should also be screened if they have risk factors like ventilation, prolonged oxygen therapy, hemodynamic instability or adverse respiratory or cardiac disease profile. This was recommended because of a number of reports of infants with higher gestation and weight developing severe ROP in India [14-16]. A particularly rapidly evolving sight-threatening form of ROP, called Aggressive Posterior ROP (APROP) has been reported in babies with higher gestations and weights in our country [16]. APROP requires immediate and aggressive management as it can quickly progress to stage 5 without passing through the classical stages. Though other unexplored factors may be responsible for this, unnecessary use of oxygen and inappropriate oxygen targets could be one of the most probable reasons [14].

NNF recommends the first screening examination for ROP should be performed not later than 4 weeks of age, irrespective of the gestational age. As a simple rule, each premature infant eligible for screening should receive the first examination by day 28 to 30 of life. Infants <28 weeks or <1200 g should be screened early at 2-3 weeks of age to enable early identification of APROP.

Programs for ROP-services in the Country

Rashtriya Bal Swasthya Karyakram (RBSK) provides child health screening and early intervention services for children, including infants up to six weeks of age. Out of the 30 pathological conditions identified under the

program, ROP and visual impairment have been included as two separate entities under 'Defects at birth' and 'Developmental Delays and Disabilities', respectively. The program entrusts the responsibility of screening up to six weeks of age with the facility providing neonatal care. Therefore, SCNUs need to integrate ROP screening with high-risk neonatal follow-up and both should be provided within or near the SCNU premises itself. This requires a trained ophthalmologist to visit SCNU follow-up clinic on a regular basis and earmarking as well as equipping a room specifically for ROP examination. In this manner, SCNU nurse can also help and assist the ophthalmologist in handling preterm infants, making the examination less stressful and safer. Under RBSK, District Early Intervention Centers can also liaison and coordinate with ophthalmologists for subsequent follow-ups. They can also organize the equipment required for screening and facilitate training of ophthalmologists. Recently, technical and operational guidelines for prevention of visual impairment and blindness from ROP and other complications of preterm birth have been released and should help in streamlining the screening and treatment process. Establishment of Zonal Mentoring Institutions at the centers of excellence for newborn care and eye care is an important step in decentralization and making services available in each district. The mentoring centers will provide technical support to operationalize ROP services, establish a monitoring system, and conduct quality assessment.

Home-Based Newborn Care (HBNC) is a national level program which caters to newborn babies and post-partum mothers. Under this program, Accredited Social Health Activist (ASHA) makes 6 to 7 home visits with the focus to identify sickness, ensure optimum growth and immunization, and ensure post-discharge follow-up of sick babies. Till now, there was no special focus on ROP. Now it has been proposed that ASHA can be a crucial community link for identification of at-risk infants as well as ensuring follow-up of such infants for ROP-screening. Realizing the difficulties in knowing the exact gestational age in many cases and to make it simple, it has been proposed that all infants <2000 g admitted to SCNU for any illness would be eligible for screening for ROP. This will make it easier for the ASHA to identify at-risk infants in the community as well as ensure timely diagnosis of APROP. Subsequently, as epidemiological data is collected and experience is gained, the criteria can be modified as required.

Over last several years, National Program for Control of Blindness (NPCB) has been spreading awareness at national level and helping in training of ophthalmologists by supporting CME and workshops,

and capacity-building of eye centers. NPCB also partners with various non-government organizations (NGO) working in field of blindness prevention. A 'National ROP Task Force' has been formed under NPCB and Ministry of Health. It is committed to developing, supporting and learning from model ROP programs in few states, implementing strategies to increase public awareness and to support parents, advocacy with government regarding the accreditation of neonatal units, exploration of role of new technology (*e.g.* new cameras, an on-line reading center for image analysis) and operational research.

There is a need for national policy and laws regarding availability of screening services for ROP in all facilities where premature or high risk neonates are managed. These may include mandatory screening policies, availability of necessary equipment and personnel trained in ROP detection and referral services. The original SCNU, toolkit which guides the establishment of new SCNUs in the country, did not specifically mention ROP screening as an essential requirement. This should now be urgently incorporated as a mandatory service in the SCNU toolkit. The updated NNF accreditation criteria for level II and III units now specifically mention ROP-screening facility as an essential requirement. Upgrading all existing SCNU and ensuring such facilities in new units will be a major boost to the national aim of reducing preventable blindness in the country. The role of Public Private Partnerships and NGO can contribute a lot to success of this initiative.

The SCNU data entry forms and database should be modified to include identification of infants at risk of developing ROP, and also to track timely screening, treatment (or referral for treatment) and follow-up of eligible babies. This will also help to identify the exact burden of the problem in various parts of the country as well as risk factors specific to our population. Based on this information, future strategies can be modified.

THE ROP TEAM

A team approach to prevention, screening and treatment lays the foundation of a successful program to prevent visual impairment due to ROP. At hospital level, the team members include, but are not limited to, a neonatologist/pediatrician, neonatal nurse, ophthalmologist, obstetrician, social worker/public health nurse, parents and hospital administrator (**Fig. 1**). The dedicated team members working in collaboration with each other help to initiate, establish and sustain a successful screening program. The role of team members should be clearly defined as has been suggested in the technical and operational document of RBSK (**Fig. 2** and **3**). The role of

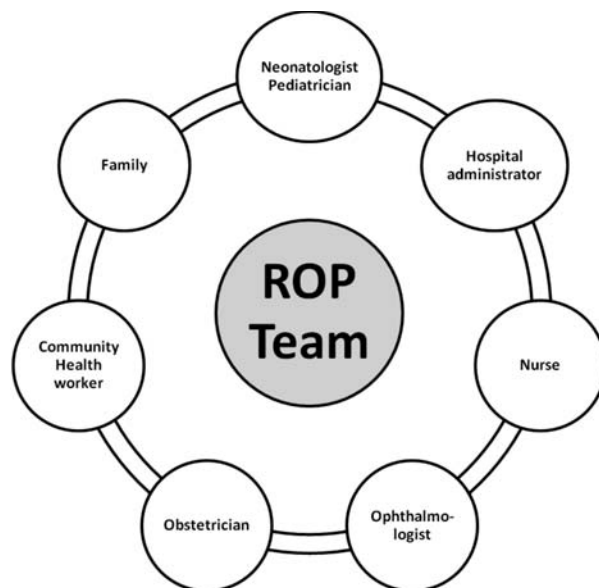


FIG.1 Composition of the ROP Team.

the pediatrician in-charge of the SNCU is summarized in **Box 1**. It is important to ensure that all team members have knowledge about relevance of the program. The following steps may help to establish a successful and sustainable program for prevention of ROP and its long term morbidities: *education* of medical/paramedical staff and parents, *early identification* of 'at risk' babies, *timely screening* of 'at risk babies', *timely treatment*, and regular *long term follow up*. The neonatologist and neonatal

Box 1 SUMMARY OF ROLES OF NEONATOLOGISTS/ PEDIATRICIANS OF CHARGE IN SNCUS

- Leadership, coordination, monitoring and evaluation
- Identify infants to be screened, ensure timely screening and follow-up, and provide support during screening
- Identify infants who missed screening/follow up and ensure they are screened urgently
- Ensure documentation of findings of screening
- Communicate findings of screening to nurses and parents
- Support ophthalmologist during treatment of ROP on the NICU, or make arrangements for discharged infants to be treated by an ophthalmologist elsewhere
- Ensure all infants screened for ROP attend District Early Intervention Centers or similar facilities for follow-up Communicate the need for further screening, if required, for all infants transferred to another neonatal unit

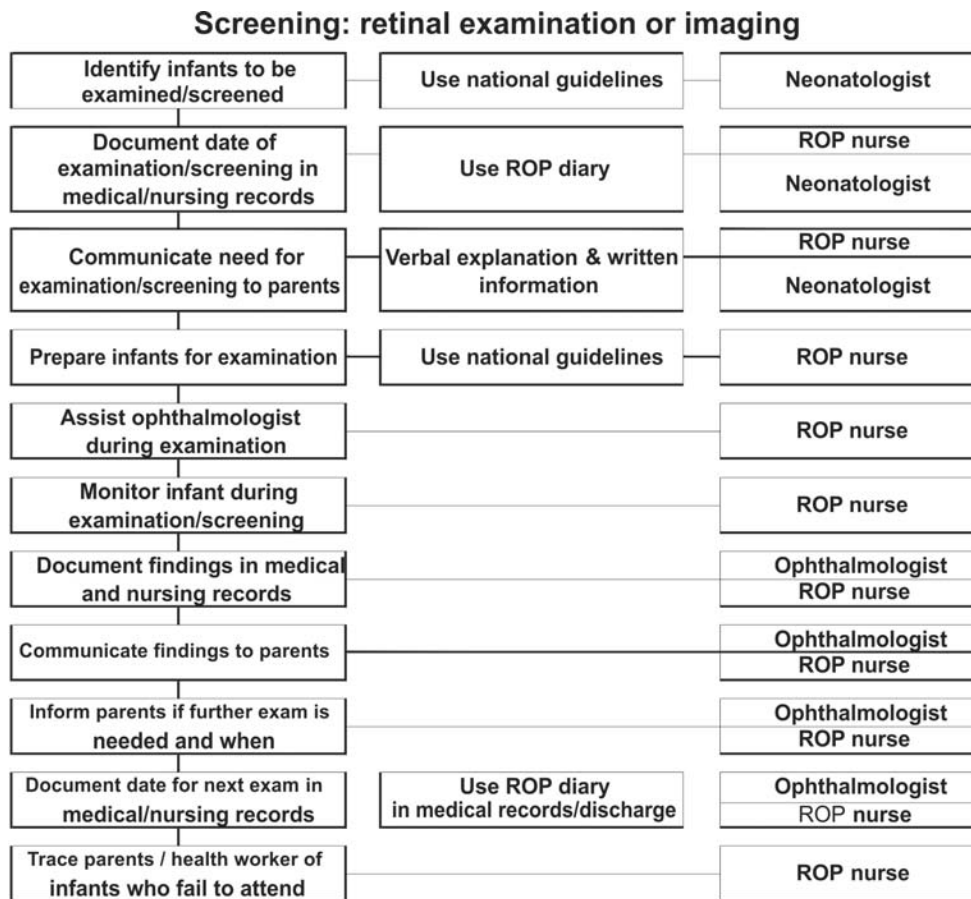


FIG. 2 Roles and responsibilities of the neonatology team and ophthalmologist (or technician) in relation to screening for ROP in the SCNU (Courtesy : Dr Rajan Shukla, Indian Institute of Public Health, Hyderabad).

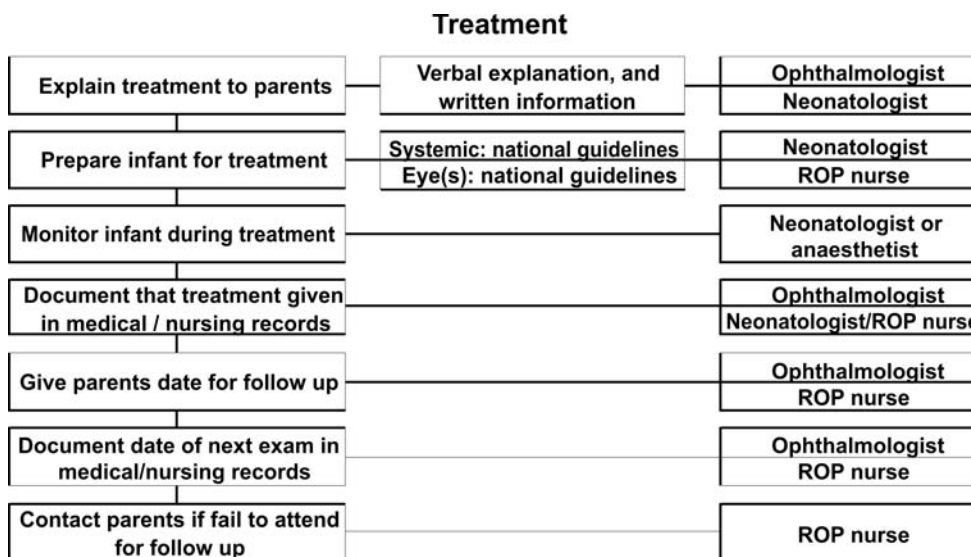


FIG. 3 Roles and responsibilities of the neonatology team and ophthalmologist in relation to treatment for ROP (Courtesy : Dr Rajan Shukla, Indian Institute of Public Health, Hyderabad)

nurse play an important role in preventing ROP by implementing evidence-based 'potentially better practices' in the hospital which include strict pre-defined criteria for oxygen saturation targets, implementation of asepsis routines, improved nutrition, restrictive blood transfusion policies etc. The formulation of such policies requires a combined effort of the neonatologist, ophthalmologist, and the hospital administration. The neonatologist and ophthalmologist play an important role by formulating other policies regarding 'whom to screen' and 'when to screen'. They, along with the hospital administration, should earmark the area of the neonatal unit where screening should be conducted. One bed in neonatal unit may be marked for care of post-treatment infants.

STRATEGIES AND TIPS FOR ROP-SCREENING PROGRAMS

In many units, one day of week is fixed as 'ROP-screening day' when the ophthalmologist screens all eligible babies identified by neonatologist/nurse. A wall chart regarding whom to screen, when to screen and how to dilate the pupil should be placed in the NICU area, nursery and the out-patient department. Whenever a baby with risk factor(s) is born or if develops a risk factor for development of ROP, the baby should be tagged and a date of ROP screening must be decided then and there. This must be followed by counseling of parents as well, regarding the presence of risk factors and the expected date of screening. A pre-printed information pamphlet in simple and local language can be utilized for educating parents about ROP and its importance. The babies identified as at-risk could be marked with color coded wrist bands or colored stickers applied on their files/cots for easy identification and as a reminder to the treating team. Vinekar and colleagues have demonstrated that screening rates can be improved even in poorly-resourced areas by low-cost red-alert cards attached to the cots of preterm infants on the day of admission [17]. The prescription and administration of appropriate drugs (e.g. eye drops for dilation) and measures to reduce pain during the process of screening and treatment is an important job responsibility of the neonatologist and the neonatal nurse. One should always remember that errors in dilution of dilating drops can prove fatal for the baby. The concentration required and the methods of dilution should be well known to the nursing staff. Charts showing the correct instructions and the required dilution of the drugs must be displayed at suitable places in the unit to prevent the medication errors. Monitoring during screening, treatment and after the treatment is another important role taken up by the neonatal nurses. The coordination between neonatologist, staff nurses and the ophthalmologist is crucial in this regard.

The ophthalmologist's role is also important for ensuring timely screening, appropriate reporting and arranging the facilities for follow up of the patients in their out-patient department (OPD). The OPDs should be well equipped to facilitate the screening. Such babies should be examined in 'special clinics' with dedicated timings. The days and timings for follow-up must be planned in such a way that parents can club the follow-up in the neonatal clinic and eye clinics on the same day. A 'ROP calendar' can be prepared and a copy also given to the family. The OPD must have a 'baby friendly' room where the environmental temperature can be maintained in the thermo-neural comfort zone for the babies. Moreover, a room in the OPD should be earmarked for feeding. Since complications like apnea and transient desaturations are known to occur with the process of screening, the ROP clinics must be well equipped to monitor such infants i.e. availability of pulse oximeters and basic training of staff in terms of suction and giving oxygen. Public health nurses or community health workers play an indispensable part by ensuring the involvement of the family in the care of the baby, counseling the family, paying home visits, tracking the patient after discharge, and contacting the families who are lost to follow up. The parents can play a very important role by ensuring the screening and regular follow-up and timely treatment of the babies.

The first ROP-screening may be performed in the NICU/SNCU under the supervision of the neonatologist. Infants who are unstable and cannot be brought out of the incubator can be safely screened as well as treated within the incubator through its sloping wall without disturbing the equilibrium [18]. The screening can be done without sedation under topical anesthesia. Comfort care techniques (e.g. administering sucrose solution, nesting, swaddling and/or the use of a pacifier) during the screening examination may be considered [19]. An ophthalmologist, who could be a pediatric ophthalmologist, a retina specialist or a general ophthalmologist well versed with indirect ophthalmoscopy, should do the examination. The person doing the procedure must have the patience and gentleness required to handle a sick and fragile newborn. Laser treatment must be performed under the supervision of a neonatologist or anesthesiologist. It is done either in NICU or in an operating room equipped with drugs and instruments required for resuscitation in the rare event of apnea/cardiac arrest. The neonatologist/anesthetist/NICU staff must monitor the baby until he/ she is fed and is stable. In case of babies who get transferred from one hospital to another, effective communication between the two hospitals is of utmost importance in order to avoid missing the 'at risk'

cases. The neonatal team must ensure long-term visual assessments during the follow up of these high risk infants as they are prone to develop refractive errors, strabismus, anisometropia and amblyopia.

One of the bottlenecks for expanding ROP-screening services is shortage of ophthalmologists trained and interested in ROP-screening. Wide-field retinal imaging using Ret-Cam by non-physicians has been shown to be effective in the KIDROP program [20]. It was shown that trained technicians using Ret-Cam for documenting disease and deciding treatment and referral, had 95.7% sensitivity, 93.2% specificity and a positive predictive value of 81.5% [20]. However, huge costs limit the widespread applicability of this model. It has been explored whether neonatologists themselves could screen for ROP. Neonatologists can screen at-risk neonates by direct ophthalmoscopy. They can independently and reliably detect posterior pole changes in babies with ROP. However, the inability of direct ophthalmoscopy to visualize the peripheral retina limits the potential of such an approach for the detailed screening. Efforts are on-going to develop new lower-cost, wide-field imaging systems which can be used widely by non-ophthalmologists. The images captured using such devices can also be sent to the experts for further opinion as well as planning the treatment. An iPhone combined with a 20-D lens has been shown to be useful in screening for ROP in resource-poor settings [21]. The subject of neonatologist-led ROP screening and its implications have been recently comprehensively reviewed [22], and we can expect major paradigm changes in ROP-screening in the coming years. Operational research needs to be conducted about several of above aspects related to ROP, including identification of high-risk infants in community, ensuring their follow-up, best models of re-training the existing ophthalmologists for ROP screening, and validation of pediatrician- or nurse-based screening.

There is also a role of collaborative learning and formation of registries and databases at the level of individual hospitals as well as at the national level. The possible benefits of collaborative learning and data-sharing in this direction cannot be overemphasized. This requires collaboration between the various institutes and administrative bodies consisting of various members of the neonatal team from different streams.

CONCLUSION

Although awareness and efforts to establish ROP-screening and treatment facilities are being scaled up in the country, there are tremendous challenges. With rapid increase in number of SCNU, the number of surviving

preterm infants needing screening and treatment for ROP is increasing exponentially. This has been rightly identified as a area of top priority by Government of India, and both neonatologists and ophthalmologists have taken up this challenge in unison to find innovative solutions suitable for our country. For the first time, there is a visible coordination between various agencies and stakeholders. With the recent activities, increasing awareness, commitment of government and the willingness of apex institutes to mentor regional and district-level hospitals, a significant decline in the incidence of preventable blindness in India can be expected in the near future.

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