

# RED REFLEX TEST (RRT) IN NEWBORN: Review article

Kshitij Aviraj Singh<sup>1</sup>, Amar Taksande<sup>1</sup>, Punam Uike<sup>1</sup>, Revat Meshram<sup>1</sup>

Institution E 147, SECTOR 2, HEC COLONY, DHURWA, RANCHI<sup>1</sup>



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## ABSTRACT

RER test is a simple, non-invasive method which can be easily done by pediatricians during neonatal clinical examination and in infants and children. Finding consistent with abnormal reflexes will point in direction of ocular pathologies. These pathologies can have severe effect on patient's vision, cognitive function and morbidity. There are many causes of preventable blindness in childhood such as congenital cataract, High refractive error, Strabismus, Congenital glaucoma retinal abnormalities like retinoblastoma, Retinopathy of prematurity. For all the above-mentioned ocular abnormalities Red Eye Reflex test plays key screening tool for their early screening and prevention of disease progression with high sensitivity. In all neonatal and pediatric departments there is a need for implementation of RRT. Proper education of pediatricians is needed in this field for prompt screening of neonates and infants, in accordance to the current guidelines.



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## 1. Introduction

The evaluation of RRT is a low-cost easy to perform skill that can be performed by pediatricians. Red eye reflex (RER) test is an effective screening tool in the early diagnosis of neonatal ocular abnormalities. In the recommendation suggested by The American Academy of Pediatrics, RER assessment should be done in neonatal period. This will not only help in early diagnosis of the ocular pathologies but will also improve the outcome with early and prompt treatment so that long term sequelae can be precluded [1]. The Red Reflex refers to the reddish-orange reflection of light which is seen coming from the back of the eye, while using an ophthalmoscope. In terms of technicality, RER is an easy to perform, simple test which is non-invasive and requires minimal equipment and settings. RER works on the principle of reflection of light. When an external light source is focused on the back of eye, it will first pass through the pupil through the pupil and the media, and then will get reflected back from the back surface of the eye and is again then transmitted through the transparent media and the pupil. This reflection is viewed as reddish-orange reflection when observed through the aperture of an Ophthalmoscope. For pathologies like cataracts, glaucoma and retinal abnormalities it is a very efficient screening tool as any factor that hinders the transmission of light rays through the transparent optical media or its reflection back from the fundus will produce an abnormal Red Eye reflex. It is of paramount importance that early screening for these ocular pathologies should be done because if these eye related pathologies remain undiagnosed and untreated, they will have their persistence into adolescence and adult life with probable permanent vision loss.

## **2. Red Reflex Technique**

Technique for RER test is a simple one. Firstly, the functionality of the ophthalmoscope checked. Room is kept dark so that the pupils are maximally dilated. Test should be performed within one week of life. A direct ophthalmoscope is to be used. The power of the lens should be kept at '0'. The eyes of neonate should be open. Voluntary eye opening is always preferred. In newborns, it is a challenging task to keep the eyes open. For this purpose the "Chair Hold" method can be used for voluntary neonatal eye opening. The examiner should hold the ophthalmoscope close to his/her eyes at distance of about 45cm from the neonate. Each eye is focused individually to see the fundus and its reflection. Examiner views both the eyes simultaneously, and then individually. For a normal RER test the fundal glow in each eye should be round, bright red to yellow in colour and the RER of both the eyes should be symmetrical. RER works on the principle of reflection of light. When an external light source is focused on the back of eye, it will first pass through the pupil through the pupil and the media, and then will get reflect back from the back surface of the eye and is again then transmitted through the transparent media and the pupil. This reflection is viewed as reddish-orange reflection when observed through the aperture of an Ophthalmoscope. An abnormal Red Eye reflex will be produced if the transmission or reflexion of light is obscured through the optical media or surface. Abnormal RER will be viewed as dull reflex, blunted reflex, dark spots, absence of the reflex or presence of a white reflex. All neonates underwent a repeat ophthalmic examination by an Ophthalmologist. Our examination findings were correlated with respect to ocular findings determined by ophthalmologist. This was followed by determining the efficacy, sensitivity and specificity of RER test in detecting neonatal ocular abnormalities. Oblique viewing technique was used to visualize both temporal and nasal retina [1], [2]. It is important for the examiner to be patient as hasty examination may result in missed diagnosis and lifetime sequelae associated with the pathology.

## **3. INTERPRETATION**

A normal RER requires a prerequisite of a clear ocular media and normal ocular surface. The ocular media consists of cornea, aqueous humor, lens, vitreous body. A normal RER test should be bright and symmetrical. Any hindrance in the anatomical path to and from the fundus or retina will produce an abnormal RER. Normally, the RER is bright Reddish-orange, but depending upon the ethnicity and skin complexion, a child may have a normal red reflex ranging from yellow to red to orange. In conditions with these colour variations, the fundal examination of the mother or father should be used for comparison. Black reflex may be caused by corneal scar, cataract or intraocular hemorrhage. A white RER is known as Leukocoria. Presence of leukocoria may point towards the diagnosis of diseases like congenital cataract, retinoblastoma and Coat's disease. Coat's disease is presence of telangetic neovascularisation of retinal vessels and the condition may be grievous for visual acuity. RER test also provides information regarding vision alignment and symmetry. In anisometric patient, patient have error of refraction and RER test may show brighter reflection in one eye as compared to other. An abnormal RER may point towards different ocular structural pathologies like the keratitis, corneal abrasion and cornea scar in the Cornea, vitreal hemorrhage or uveitis in vitreous. In the examined cases, we have taken into account parameters and risk factors which include features like weight at birth, maternal gestational age at delivery, oxygen therapy, transfusion of blood product and phototherapy were evaluated which could effect visual acuity and RER test results [3].

## **4. Reduced or Absent RRT in Newborn**

Reduced RER, White RER or absent RER should be considered as a medical emergency in a neonate. In the neonatal congenital cataract is the most common cause for white RER [4]. With white RER, the diagnosis of Retinoblastoma should be kept in mind as if the disease remain unattended, it may prove to be fatal [5]. Early screening and prompt action may prevent sequela associated with the underlying pathologies.

## Retinoblastoma

Retinoblastoma is the most common intraocular malignancy in childhood, with an incidence of approximately 1:20000 live births per year, which leads to blindness and is life-threatening if left untreated. With 60% of it being unilateral and non heritable, it covers up seventeen percent of the neoplasm in the neonates. Its onset can occur in utero to up to 4 years of age [6], [7]. Heritable forms of retinoblastoma are usually present in under five population, are small and bilateral. Following surgical treatment, they good outcome in visual acuity. A patient with positive family history of retinoblastoma should be thoroughly examined by an expert ophthalmologist even in presence of a normal RER test and should be followed regularly till 3 years of age. Most common symptoms include leukocoria evident as white eyes on photographs taken with flashlight. Patient may also present with strabismus, heterochromia, hyphema and signs of ocular inflammation like redness, itching, pain or tears [8- 10]. In current scenario, RER Test is considered the best screening tool for detection of retinoblastoma. Though a white RER points towards the diagnosis of retinoblastoma, presence of a normal RER should not be used to rule out retinoblastoma. Early screening of retinoblastoma is important as it is a fast growing invasive tumor with metastatic properties which should be early detected and promptly treated.

## 5. Congenital cataract

The lens opacification at time of birth or within one year of age is termed as congenital cataract. The occurrence of congenital cataract is between 0.6 to 15 in every 10000 live births. Congenital cataract is estimated to be responsible for one tenth of total childhood blindness [12- 14]. Most of the unilateral cataract are idiopathic with no syndromic association however, upto one fifth of cataracts are associated with congenital rubella. Most common causes of congenital cataract includes chromosomal abnormalities (trisomy 21, trisomy 18), metabolic disorders (galactosaemia) and congenital infection syndrome (toxoplasmosis, cytomegalovirus, syphilis, rubella, herpes simplex virus, varicella zoster virus). Although bilateral cataracts are not associated with genetic mutations but they need further evaluation to exclude systemic association. Early detection and treatment of congenital cataract has become a priority of the Global Vision 2020 initiatives of the World Health Organization. The RRT serves as a highly sensitive screening test for congenital cataract. [15] reported congenital cataract incidence of 0.009% while screening of neonates using RRT. [16] reported the higher sensitivity of RRT in comparison to other diagnostic techniques in the detection of congenital cataract. Lack of red reflex during the routine neonatal eye screening is one of the earliest indicators of a child with congenital cataract. [17] reported a 5-month infant with bilateral congenital cataract, who was investigated for rubella and galactosemia at time of birth. But later he presented with progressive worsening systemic condition including delayed motor milestones, generalized motor weakness, supraventricular tachycardia, with positive family history of neonatal deaths, & detectable lactic acidosis and hypertrophic cardiomyopathy leading to the diagnosis of Senger's disease. Other associated signs with congenital cataract include nystagmus, absence of interest in the surroundings and inability to fix and follow objects. Early detection of this condition is crucial, since early therapeutic intervention before the age of 6 wk for unilateral cases and 8 wk for bilateral cataracts are associated with best visual outcome. Delay in diagnosis or treatment can lead to poor visual gain due to development of amblyopia which has negative impact on the neurobiological development of the children. [12] reported that more than half of the infants with isolated cataract were diagnosed during the first 6 wk of life, while 38% of them were detected later. These results underlined the role of screening methods in early detection of such cases.

## 6. Retinopathy of prematurity

Amongst various causes of blindness worldwide Retinopathy of Prematurity forms a major one. Retinopathy of prematurity (ROP) is a vasoproliferative disorder of retina that predominantly affects

premature babies, especially those with low birth weight. [18] reported ROP as one of the commonest cause of leukocoria in infants, contributing more than 10% of all cases of leukocoria. Gestational age of less than 34 weeks, birth weight less than 2000 g, history of oxygen therapy, septicemia and blood transfusion are common risk factors for the development of this potentially blinding disease. Since year 1940 the world has seen three epidemics of Retinopathy of Prematurity. The Stages of ROP is presence of the demarcation line, (Stage 1) at the junction of vascularized and avascular retina. As the disease progresses, capillary growth begins at the edges of the demarcation line leading to the formation of "ridge" (Stage 2); with further proliferation of fibrovascular tissue from the ridge into the vitreous body which is categorized as stage 3. With further worsening these fibrovascular membrane contracts causing tractions at the retina leading to partial detachment (Stage 4) or total detachment of the retina (Stage 5). Advanced stages have very poor surgical outcome making it very important to have robust screening method for careful examination of retina in all preterm infants by a pediatric ophthalmologist, screening should be at 4 weeks postnatally and has to be emphasized and encouraged by pediatricians. As Severity of this disease greatly depends on the varying level of care in the community. It is worth noting that leukocoria may be detected after Stage 3 of ROP, however, the RRT performed may help in early detection & referral of all suspicious cases. Refractive errors, amblyopia and strabismus more common in children with ROP.

## **7. ADVANTAGES OF RRT**

RER test offers multiple advantages. Firstly, it is a very easy procedure to perform. The equipments required for the test are minimal, thus making it very feasible and the setup required are also very simple thus making it cost effective. Considering the burden of neonatal ocular pathologies and the ease of doing RER test and the multiple study results showing its efficacy and high sensitivity and specificity, it is very easy to conclude that RER test is the best tool in current day scenario for neonatal and infantile ocular abnormality screening. Moreover, the skill can be easily learned and only takes minutes to perform. The large number of false positive test gets outweighed by the fact that grievous complications associated with the positive pathological findings and improved outcomes associated with timely interventions.

## **8. LIMITATIONS OF RRT**

Technically, the RER test is a simple, non-invasive, very easy to perform & requiring minimal settings and equipment. But for the test to commence, the neonate's eyes should be open and neonate should be relaxed. Voluntary opening of the eyes of the neonates causes increased irritability and thus interferes with the examination results.

Repeat detailed examination by the ophthalmologist requires speculum application for lid retraction. This process caused increased irritability and cry in the neonates. Thus, such an examination required presence of a paediatrician and SOS NICU transfer.

## **9. ALTERNATIVE METHODS TO RRT**

World Health Organization (WHO) in their recent recommendation included pencil light examination for neonatal ocular screening while refrained from RER test for screening of ocular abnormalities for newborn. Study by [19] gave strong recommendation regarding inclusion of RER test and exclusion of torch light examination in the WHO guidelines by showing low sensitivity of torchlight for neonatal ocular abnormality screening. A study performed in Turkey [20] showed that congenital cataract diagnosis was missed on most occasions when neonates were screened by torch light examination. While, there are multiple studies who support the efficacy, sensitivity and specificity of RER test in neonatal ocular pathology screening.

## 10. Recent Studies Related To RRT in Newborn

[21] in his study for Brückner test found that RER test to be an effective screening tool for neonatal and infantile ocular defects, binocular vision and problems of visual acuity. [15] in his study screened 22,272 neonates with Red Reflex Test in a study period of 3 years from 2012-14. In the screening 461 neonates (4.83%) came out with an equivocal or abnormal red eye reflex, who were later subjected to detailed ophthalmic examination by Ophthalmologist in specialized centres. Of these later subjects, retinoblastoma in two neonates and congenital cataract in one was found. Followup study showed didn't show any congenital anomalies in the subjects with normal red eye reflex, hence proving the test to be highly sensitive. Study by [22] incidence of abnormal RER was increased in neonates born to mother with difficult or prolonged labour. Study by The [23] showed that RER test screening is more effective for screening of anterior segment defects as compared to posterior segment [24] in their study screened 15,398 neonate, out of which 1266 subjects had abnormal or equivocal test results which on further screening found twelve different set of ocular abnormalities.

## 11. Conclusion

RER test should be incorporated as an essential tool for screening of neonates for ocular anomalies as a regular practice. Multiple studies have supported this test to be highly sensitive, specific and not only the test is very feasible and low cost, it can be performed very easily with minimal equipments and setting required. Early detection and prompt and effective treatment of neonatal ocular anomalies is important as it will help us to prevent long term sequale associated with the disease consideration and at the same time help in improving the quality of life.

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