

The Red Reflex Examination in Neonates: An Efficient Tool for Early Diagnosis of Congenital Ocular Diseases

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ABSTRACT: **Background:** The American Academy of Pediatrics recently published recommendations for the red reflex assessment in the newborn period to detect and treat ocular disorders as early as possible, and to prevent lifelong visual impairment and even save lives. The test is technically simple to perform, non-invasive, requires minimal equipment and can detect a variety of ocular pathologies including cataracts and retinal abnormalities. No specific national guidelines exist on this issue.

Objectives: To document the implementation of red reflex examination in routine neonatal care and present the findings.

Methods: Our clinical experience following inclusion of the red reflex test into the newborn physical examination in a single center was reviewed. In addition, an electronic mail questionnaire was sent to all neonatology departments in Israel regarding performance of the red reflex test.

Results: During 2007–2008, five infants were identified with congenital cataracts at days 2–6 of life prior to discharge from hospital. Surgery was performed in one infant at age 2 months and all infants underwent a thorough follow-up. The incidence of congenital cataract in our center was 1:2300. Less than half the neonatology departments have endorsed the AAP recommendation and perform the red reflex test routinely.

Conclusions: Abnormal red reflex test after delivery enables a rapid ophthalmologic diagnosis, intervention and close follow-up. We recommend that red reflex screening be performed as part of the newborn physical examination; if abnormal, an urgent ophthalmologic referral should be made.

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KEY WORDS: red reflex, visual screening, congenital cataract, retina, newborn

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The American Academy of Pediatrics has recently published recommendations that the red reflex assessment be conducted in the newborn period in order to detect and treat ocular disorders as early as possible, and to prevent lifelong visual

impairment and even save lives [1-3]. Congenital cataract diagnosis and treatment have become a priority of the Global Vision 2020 initiative of the World Health Organization [4] as they remain the most common treatable cause of visual disability in infancy and childhood [5]. Untreated congenital cataracts are responsible for approximately 10% of all childhood blindness worldwide [5,6]. The incidence of childhood blindness due to congenital causes has been reported in western countries as 1–6 per 10,000 live births; etiologies include intrauterine infections, metabolic disorders, chromosomal abnormalities and inheritance [5,7].

In contrast to adult age-related cataract where the surgical intervention may be delayed without affecting the visual outcome, the timing of cataract removal in infancy is critical to attain maximal visual potential after surgery [8]. This fact has been related to the unique developmental physiology of the human visual system, which becomes sensitive to visual deprivation 6 weeks after birth following a latent period of visual immaturity [9]. Therefore, delayed surgery for significant congenital cataracts may result in abnormal visual development with irreversible amblyopia. To achieve favorable long-term visual outcome the recommended best practice for treatment of cataracts that obscure the visual axis includes an early surgical intervention, possibly within the first 3 months of life and as early as 6 weeks in unilateral disease [10-12].

To detect early eye pathology and institute rapid referral and treatment, universal screening of newborns using the red reflex testing is conducted in many countries, resulting in early diagnosis of various eye pathologies including not only cataracts, but also glaucoma, retinoblastoma, retinal abnormalities, systemic diseases with ocular manifestations, and high refractive errors [1-3]. Red reflex testing may be performed simply and rapidly as part of the routine physical examination.

In this report we examine the effects of implementation of the red reflex examination in our hospital over a 2 year period (2007–2008). We describe the process of implementing the screening program using the red reflex test, provide the rates of detection of ocular pathologies diagnosed following false positive results (i.e., unnecessary eye consultation and outcome of identified patients), and false negative rates (undiagnosed pathologies after birth that were later diag-

AAP = American Academy of Pediatrics

nosed as congenital eye pathology). In addition, a survey of the test's implementation in neonatology departments across Israel was performed.

PATIENTS AND METHODS

Kaplan Medical Center is a university-affiliated local general hospital in Rehovot with 5500–6800 deliveries annually. While the red reflex test has been performed sporadically by a few neonatologists for several years, in 2007 we implemented red reflex testing as a routine part of the newborn physical examination. The pediatric ophthalmologists briefly instructed neonatologists and pediatric residents.

The test is performed with a direct ophthalmoscope, set at 0 lens power, held close to the examiner's eye and focused on each pupil individually at approximately 45 cm from the eye. Both eyes are then viewed simultaneously. The red reflex seen in each eye individually should be round, bright reddish-yellow (or light gray in darkly pigmented, brown-eyed patients) and should be similar in both eyes. Dark spots in the red reflex, a blunted dull red reflex, lack of a red reflex, or presence of a white reflex are all indications for ophthalmology referral. To maximize pupil dilation the red reflex test may be preferably performed in a darkened room [1,3]. If the test cannot be performed shortly after birth, mostly due to edema of the eyelids, it is deferred to the second day of life.

A telephone or electronic mail questionnaire on the performance of the red reflex test was circulated (December 2008) to all neonatology departments in Israel.

RESULTS

During the 2 year period of the study 5 of 11,500 newborns were diagnosed with congenital cataracts, an incidence of 1:2300. The neonatal and ophthalmologic characteristics of these neonates are shown in Table 1. All cataracts were identified and diagnosed at days 2–6 of life. Infectious and metabolic evaluations were negative.

There were 12 referrals for ophthalmologic evaluation due to abnormal red reflex test, of which 5 were found to have ocu-

lar pathology, as described. Therefore, the red reflex test has a specificity of 42% during the initial implementation period, which is expected to be higher following increasing learning curve and clinical experience. The false positive rate was low: 1 in 1643 (7 of 11,500; 0.0006%). To the best of our knowledge, during 2007–2008 only one infant was subsequently diagnosed at age 8 months with cataract. The absence of prospective follow-up, however, precludes calculation of the true false negative rate, although this appears to be very low.

A survey of the 26 neonatology departments in Israel revealed that only in 12 was the red reflex eye assessment routinely performed prior to discharge of the newborns. The main reason cited for not conducting the test included limited staff and reluctance to "perform the ophthalmologist's job." All the departments that routinely perform the test have encountered approximately 1–2 cases a year of abnormal findings leading to a diagnosis of congenital cataract.

DISCUSSION

Use of the red reflex as a screening tool was originally described by Bruckner in 1962 [13]. To generate a red reflex, the light must travel directly through the clear colorless ocular media: the cornea, aqueous humor, lens and vitreous body. A normal red reflex requires clarity of each element. The reflex is not actually generated from the retina, which is transparent except for its outermost pigmented layer. Although Bruckner originally described the normal reflex as red, the normal "red reflex" is often yellow, orange, red, or any combination. An abnormal test includes its absence, asymmetry, leukocoria (white reflection from the retina), or non-homogenous reflex. The test may be abnormal when there is opacity in the ocular media such as in atrophy of the optic nerve, cataract, intraocular hemorrhage, retinal detachment or intraocular tumor, as well as in high refractive disorders, anisometropia and ocular misalignment (strabismus) [13,14].

It should be noted that high risk infants, including those with familial cataract, family history of retinoblastoma, metabolic disorders associated with ocular diseases, microphthalmia or eyelid hemangioma (port-wine stain or

Table 1. Characteristics of newborns with abnormal red light reflex examination at discharge from hospital, eye pathology and follow-up

Patient	Gestational age (wks)/birth weight (g)	Red light reflex findings	Eye examination findings	Ophthalmology treatment and follow-up
1	40/3000, female	Bilateral black dots	Bilateral small antral cataracts	Persistent size of opacity, follow-up
2	34/1900, female	Bilateral white dots	Bilateral small cataracts	Strict follow-up
3	38/2770, female	No red light reflex, right eye	Right eye congenital cataract, persistent hyperplastic primary vitreous	2 mos: aspiration of cataract and removal of vitreous 7 mos: removal of lens, pars plana vitrectomy, contact lens for prevention of amblyopia
4	38/2520, female	No red light reflex, both eyes	Microcornea, coloboma of iris, optic disc, and lens, both eyes	Follow-up
5	40/3716, male	Bilateral decrease central opacity	Bilateral persistent papillary membrane with minute anterior polar lens opacity	Follow-up

extensive capillary), should be examined by an experienced ophthalmologist.

We found that the red reflex test was simple to perform and requires little time and minimal equipment. Within a short time it has been integrated into the newborn physical examination and has detected abnormalities and prevented vision loss. Thus, we strongly recommend that red reflex testing be performed in every neonatology department.

We wish to add that since submission of this article for publication in *IMAJ*, a clinical guideline on this issue, proposed by a joint committee of the Israeli Pediatric Ophthalmology and Neonatal Societies, was approved, thereby mandating routine screening for red reflex in Israel.

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