Red Reflex Examination in Neonates: The Need for Early screening

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Children are a priority in "Vision 2020," the World Health Organization's global initiative for the prevention of avoidable visual impairment [1]. Congenital cataract is the leading cause of preventable childhood partial sight or blindness [2] – both by primary prevention, for example, through a rubella immunization program, and by secondary prevention depending on early screening of the pupillary red reflex. Given that the optimal time to remove a dense congenital cataract in an infant and initiate optical treatment appears to be age 4 to 6 weeks [3], screening to ensure prompt early treatment is essential for improving visual outcome [4]. Newborn screening for media opacities, comprising examination of red reflex, is widely accepted. The red reflex test uses transmission of light from an ophthalmoscope through all the normally transparent parts of a subject's eye. Any factor that impedes or blocks this optical pathway will result in an abnormality of the red reflex. Corneal opacities, aqueous opacities, iris abnormalities, cataracts, vitreous opacities, and retinal abnormalities including tumors or chorioretinal colobomata, may produce abnormalities or asymmetry of the red reflex [5].

Screening newborns with the red reflex test is widely accepted. The American Academy of Pediatrics [6] and the British Paediatric Association [7] currently recommend red reflex assessment as a component of the eye evaluation in the neonatal period and during all subsequent routine health supervision visits. The purpose of this policy statement is to minimize the risk of delay in the diagnosis of serious vision-threatening or life-threatening disorders. Despite these worldwide recommendations, there are large variations in the implementation of red reflex examination [8-12]. In the UK, less than half the cases in the 1995–1996 cohort of congenital and infantile cataract were detected by screening examinations at age 8 weeks or less [8]. Similarly, a recent study of infantile cataracts in the United States showed that 38% were diagnosed after age 6 weeks [9]. Reporting their 10 year experience in a single regional ophthalmology center, Sotomi et al. [10] showed that none of the 27 infants with congenital cataracts was diagnosed by the newborn screening examination. Six of 8 infants who were diagnosed before 3 months of age had a good visual outcome in contrast to only 3 of 19 diagnosed after 3 months. Considering that this practice was not evaluated prospectively, it is not clear whether the low detection rate is the result of non-compliance, inadequate technique, or low sensitivity of the red reflex test as a screening tool.

In this issue of *IMAJ*, Eventov-Friedman and colleagues [14] report a single-center clinical experience following implementation of the red reflex test as part of the newborn physical examination. During the 2 year study period, of 11,500 newborns who were screened with red reflex examination, 12 were referred to ophthalmology consultation due to suspected abnormal red reflex. In 5, the diagnosis of congenital cataract was confirmed, giving an incidence of 4.3 per 10,000 newborns. Based on routine notification systems for monitoring congenital anomalies in the USA and Europe, the current annual birth prevalence of congenital or infantile cataract has been estimated to be approximately 1 per 10,000 of the total number of births. The British Congenital Cataract Interest Group reported a cumulative incidence of congenital and infantile cataract of 2.29 per 10,000 by age 1 year [13]. In comparison to these findings, the reported incidence in the current study is higher, suggesting a high detection rate. However, in the absence of follow-up or national surveillance for the diseases, the incidence and sensitivity of red reflex have yet to be determined. The positive predictive value (42%) that was shown here, after a short period of implementation, is also higher than that shown elsewhere [15], yet over-referral is unavoidable in the effort to detect infants with congenital media opacities.

The authors included in their report a survey of all Israeli neonatal departments on the implementation of the red reflex test in the newborn examination, showing that until December 2008 only 12 of 26 neonatal departments routinely assessed the red reflex prior to discharge. The disparity between the incidence of congenital cataract in the current study and the 0.68 cases per 10,000 newborns reported to the Israeli registry of congenital anomalies during the years 2000–2008 (personal communication) should encourage neonatologists to implement this screening

for early diagnosis of eye pathology. The Israel Neonatology Association guidelines for routine red reflex examination in newborns published in July 2009 [16] constitute the first step for improving the quality of red reflex screening. However, more specific guidance regarding the purpose and content of red reflex examination and the promotion of programs for training all involved in its management are required. In addition, there is a need for repeated examination before age 6 weeks. In all infants with a family history of retinoblastoma or cataract, neurologic or metabolic disorders, and microphthalmia or eyelid hemangioma, consultation with an experienced ophthalmologist should be emphasized.

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