

Assessment of vision in paediatric primary care practice

Dunja Bajtl¹, Mirjana Bjeloš^{2,3,4}, Mladen Bušić^{2,3,4}, Ana Ćurić^{3,4}, Vladimir Bajtl⁵, Josip Barać^{1,2}

Context: Assessment of visual function often poses a challenge for a paediatrician in primary practice. However, due to the simplicity of basic visual assessment methods, paediatricians have the means to evaluate a child's visual function. As a crucial part of every preventive physical examination of a child, the red reflex test is the most important tool for discovering obstruction of the visual axis as the cause of deprivation amblyopia. The visual axis can be obstructed due to various vision - or even life-threatening conditions.

Aim: To present visual assessment methods available to the paediatrician in primary practice.

Data source: PubMed, Google Scholar, Scopus, and available Internet screening guidelines.

Conclusion: If obstruction of the visual axis is suspected, immediate referral to a paediatric ophthalmologist is mandatory.

Keywords: CHILD; VISION TESTS; VISUAL ACUITY; OPHTHALMOSCOPES; AMBLYOPIA

INTRODUCTION

Amblyopia is the most common form of visual impairment in children and the leading cause of monocular blindness in patients aged 20-70 years (1, 2). It is defined as impaired vision due to structural and functional changes in the visual cortex caused by incomplete visual experience during the first years of life (2). The estimated prevalence of amblyopia in 4-year-old children in Croatia is 8.08% (3) compared to the commonly reported 0.3% - 5% in the world (2).

The are two forms of abnormal sensory stimulation that cause amblyopia: deprivation of foveal image (unclear image) caused by the obstruction of the visual axis (deprivation amblyopia) and abnormal binocular interaction (interocular competition from two foveas) caused by strabismus or refractive errors (strabismic and refractive amblyopia) (2). Deprivation of the visual axis during the first year of life causes irreversible reorganization of the visual pathway with a predominance of cortical neurons arising from the non-deprived eye (2). Prompt identification and referral of the child are of the utmost importance for a child's visual habilitation. This paper thoroughly elucidates methods for vision screening, available in primary paediatric care practice during the first year of life, to eliminate amblyopia which otherwise leads to severe visual impairment and blindness.

EXTERNAL EXAMINATION

An inspection alone can reveal gross structural abnormalities obstructing the visual axis, tearing discharge or abnormal eye movements. Obstruction of the visual axis refers to abnormalities of the eyelids, cornea, pupil, crystalline lens, vitreous or to diseases of the choroid and retina (e.g., retinoblastoma) that prevent light from entering the eye through the pupil and projecting onto the retina in the fovea centralis. Every obstruction of the visual axis is a cause for referral.

Paediatricians should look for symmetry in a child's face. Infants at birth have a relatively narrow palpebral aperture. A

¹ University Eye Department, University Hospital Centre Osijek, Osijek, Croatia

² Faculty of Medicine, Josip Juraj Strossmayer University of Osijek, Osijek, Croatia

³ University Eye Department, Reference Centre of the Ministry of Health of the Republic of Croatia for Paediatric Ophthalmology and Strabismus, University Hospital "Sveti Duh", Zagreb, Croatia

⁴ Faculty of Dental Medicine and Health Osijek, Josip Juraj Strossmayer University of Osijek, Osijek, Croatia

⁵ University Department of Otolaryngology, University Hospital Centre Osijek, Osijek, Croatia

Correspondence to:

Assoc. Prof. Mirjana Bjeloš, MD, PhD, Reference Centre of the Ministry of Health of the Republic of Croatia for paediatric ophthalmology and strabismus, University Eye Department, University Hospital "Sveti Duh", Sveti Duh 64, 10 000 Zagreb, Croatia,
e-mail: dr.mbjelos@gmail.com

normal horizontal corneal diameter (white-to-white, WTW) at term birth is 9–10.5 mm (5). Macrocornea is defined as a WTW distance of 11 mm or more in term infants (5). A corneal diameter of less than 9 mm is a sign of microcornea (5).

Pupillary response to light should be noted using a bright light to examine each pupil for direct and consensual response in both photopic and scotopic conditions (6).

Pupillary light reflex is absent in premature infants at 30 weeks of gestational age (GD) and is well developed by 35 weeks of GD (7).

Tear production in children is similar to that in adults (5). The normal tear meniscus height in children is approximately 0.2 mm (8). The height of the tear meniscus can be used to measure tear volume. The elevated tear meniscus height indicates the existence of a nasolacrimal duct obstruction, a condition especially important in infants (5).

Eye discharge or excessive tearing from one or both eyes may indicate a pathologic condition. Purulent discharge can occur as a result of bacterial infection (e.g. *Neisseria gonorrhoeae*) or a chemical irritant. Abundant tearing may result from a blocked tear duct, foreign body, allergic reaction or infection (9).

The classic triad of tearing, photophobia and blepharospasm indicates congenital glaucoma, usually appearing during the first three months of life (10). Congenital glaucoma is a vision-threatening condition and, as such, is an urgent cause for referral. Corneal clouding and oedema with conjunctival injection are often associated signs.

HIRSCHBERG TEST

This simple test objectively determines the eye position depending on the corneal light reflection position while simultaneously lighting both eyes (2). The subject is instructed to look at the tip of a penlight. Two positions are assessed: decentration of the corneal light reflex relative to the center of the pupil and the position of the corneal light reflex in one eye relative to the other eye. A 1 mm of corneal light reflection shift away from the center of the pupil corresponds to a deviation of 7° or 15 prism dioptres (PD) (2).

RED REFLEX

The red reflex (Brückner) test is a vital part of neonatal, infant and every preventive physical examination of a child (11, 12). It is essential for the early detection of vision- and life-threatening conditions that cause deprivation amblyopia, such as cataracts, retinoblastoma, corneal opacities and retinal abnormalities (12). The red reflex test uses the transmission of light from the direct ophthalmoscope (12).

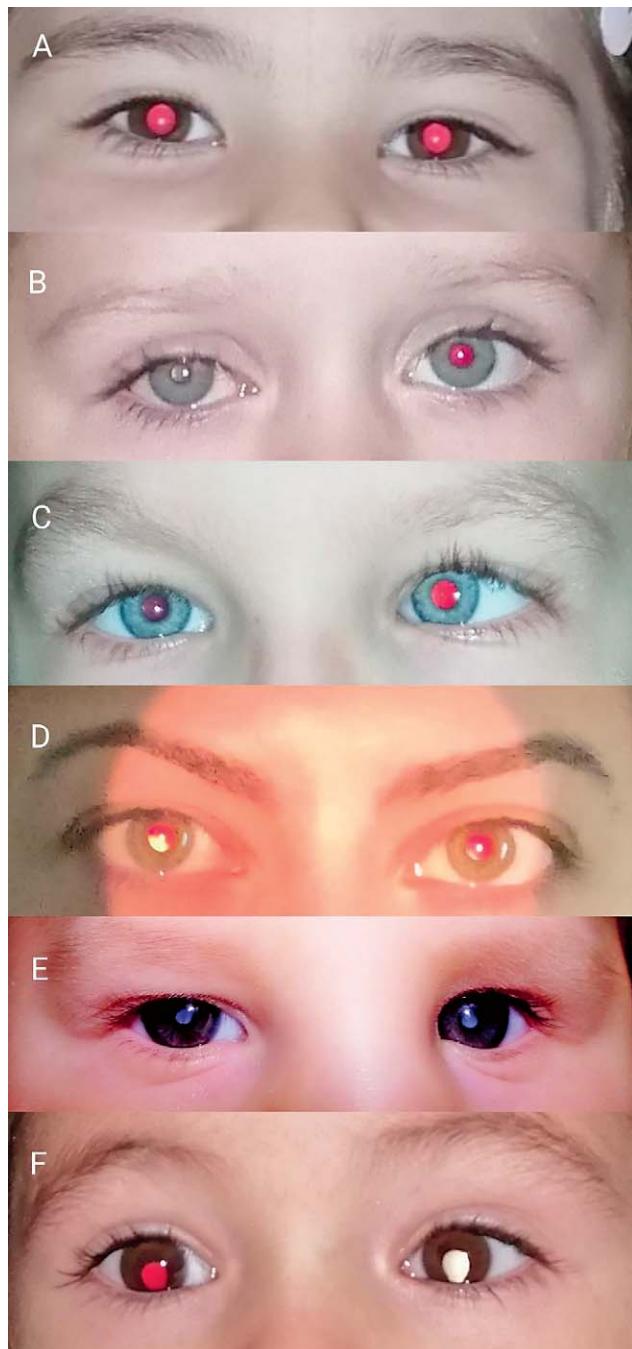


FIGURE 1. Red reflex. (A) Healthy eye. (B) Right eye leukocoria due to retinoblastoma. (C) Left eye esotropia. (D) Myopia, with a higher grade of myopia in the right eye. (E) Bilateral leukocoria due to cataracts. (F) Bilateral iris colobomas with left leukocoria due to large chorioretinal coloboma. (Source: authors' archive)

According to the new Plan and Programme of Health Care Measures for the period 2020–2022 issued by the Ministry of Health of the Republic of Croatia (NN 142/2020-2753), the red reflex test is a mandatory part of the newborn examination in maternity wards performed by a paediatrician, subspecialist neonatologist/subspecialist in the field of paediatric intensive care medicine, and a part of a general medi-

cal examination for infants who are 1-2 months old performed by a primary care paediatrician (13). A red reflex examination is recommended by paediatric societies across the world (11, 14, 15, 16).

Performing the examination requires dimming the lights in the examination room to allow the pupils to dilate. If the pupils are not large enough to allow light to enter the eye, it will result in an absent reflex.

The red reflex test is performed using the largest light diameter of the beam with the lens set to bring the eyes into focus. The light from the ophthalmoscope should be simultaneously projected onto both eyes at a distance of approximately 1 meter and then each eye separately at a distance of 40 cm (2, 17, 18). The presence of asymmetry for the red reflex in terms of color, brightness and size should be determined binocularly (Figure 1) (2).

Normal red reflex. A normal result means that the red reflexes are symmetric, red, orange, or yellow (19) and occur concurrently in both eyes (Figure 1A).

Abnormal red reflex. Any asymmetry requires an urgent examination by a paediatric ophthalmologist. An absent, dark or white (leukocoria) reflex indicates an obstruction that prevents light from reflecting to the physician's eye and can occur due to cataract, corneal scarring, vitreous hemorrhage, retinal or optic disc abnormalities (Figure 1B-F) (7, 20, 21).

Retinoblastoma is the most common primary intraocular malignancy and the seventh most common malignancy in childhood (Figure 1B) (7, 20). Not to be confused with the previous description, the optic disc can manifest as pseudo-leukocoria in certain gaze positions.

Paediatric cataract. Paediatric cataracts are the leading cause of treatable blindness in children. The overall prevalence of childhood cataracts and congenital cataracts is 0.32 to 22.9/10000 children and 0.63-9.74/10000 children, respectively (23). Overall, the incidence ranges from 1.8 to 3.6/10000 children per year (23). Congenital cataracts are hereditary in 8.3%-25% of cases, with 75% attributed to autosomal dominant disorder (24). Congenital cataracts are associated with congenital infections. The TORCH syndrome refers to an infection in a developing fetus or newborn caused by any group of infectious agents. The rubella virus is the most common causative infectious agent for non-hereditary paediatric cataracts (17). Other ocular or systemic abnormalities are often present in children with congenital cataracts. Depending on the density and localization of the opacity, the reflex can be absent, white or irregular (Figure 1E). Surgery is advised in visually significant opacities if more than 3 mm of central opacity is present. In such cases, cataract surgery is advised to be performed at the age of

6-8 weeks (2). The advent of nystagmus indicates severe, irreversible visual damage (25).

Strabismus. Strabismus, an important amblyogenic condition, causes a brighter red reflex from the deviated eye (Figure 1C). Importantly, strabismus may also be a sign of retinoblastoma (2). Paediatricians are not expected to anticipate the diagnosis behind an abnormal red reflex; however, immediate referral to an ophthalmologist is mandatory (12).

Anisometropia. The red reflex test is considered useful in detecting anisometropia (19). The inter-ocular difference in brightness is strongly suggestive of anisometropia (Figure 1D) (19).

Pseudostrabismus. Pseudostrabismus is a condition when eyes are well aligned as defined by the cover test; however, they appear to be misaligned (19). It is often a cause for an unfounded referral of an infant to a paediatric ophthalmologist, usually due to epicanthal folds or a broad nasal bridge. Parents usually report less sclera is visible nasally in the eye assumed to be squinting compared to the healthy eye (20). This should never guide the paediatrician in establishing a diagnosis for strabismus. Such cases are clearly differentiated by red reflex testing.

ASSESSMENT OF VISION

Fixation is an indispensable reflex for visual development. The ability to steadily fixate is usually acquired during the third month of life due to the maturation of the fovea and the central nervous system (CNS) (26). Newborns demonstrate the largest fixation preference for human faces. Fixation should not be examined using a flashlight. The child should maintain fixation while fixating with both eyes and monocularly. Performing an observation of symmetry in the child's behaviour while fixating monocularly is very important in assessing the vision of infants. The inability to maintain fixation with only one eye, with the opposite eye covered (occluded), or rejecting occlusion of only one eye, is presumptive evidence of a difference in the quality of vision between the two eyes.

Children as young as six weeks should have some response to a physician's face for a close view at a distance of 10-20 cm (27). The normal development of vision is presented in Table 1.

If binocular fixating and following are not evident by three months of age, a referral is recommended (28). Poor fixation can be used as a sign of both CNS lesions and ocular disorders (29).

VESTIBULAR-OCULAR REFLEX EXAMINATION (DOLL'S HEAD MANEUVER)

The doll's head maneuver is primarily used for neurological examination of the 3rd, 6th and 8th cranial nerves, the functioning of its nucleus in the *medulla oblongata*, and for assessing the functioning of the extraocular muscles. The vestibular-ocular reflex (VOR) provides foveal alignment on an object of attention during brief, high-frequency head movements by generating ocular rotations of equal speed in the opposite direction, such as head movements (30). To elicit VOR, the child's head is briskly turned from side to side and reaches the end position at each turn (30). Normally, if the head is rotated 10°, the eyes will rotate exactly 10° in the opposite direction, equally fast, to maintain the foveation of a stationary target (30). Any imbalance in the VOR results in the eyes being dragged off the target at the end of the head thrust, followed by a refixation saccade to regain the fixation. Eye movement abnormalities during VOR examination can be caused by either peripheral (otolith dysfunction) or central disruptions of vestibular activity (lesions of the brainstem or cerebellum) (30, 31). The test can be performed as early as 25 weeks of gestational age (30).

COVER TESTING

Cover testing is the most commonly used method for determining the presence, type, and amount of ocular misalignment. Cover testing should be performed near (an accommodative target held at a distance of 33 cm for fixation) and at a distance (of 3-5 m) (32). This test is feasible upon stabilizing the fixational reflex and conjugate eye movements (Table 1).

Importantly, paediatricians should differentiate between the cover and uncover testing. Such testing is always performed simultaneously (2). The actual test is performed by using an opaque or fogged occluder (or any opaque object of adequate size and shape to occlude the entire eye, for example, the physician's palm) to occlude the healthy, fixating eye. Hence, in cover testing, the apparently fixating eye is covered by the occluder. During occlusion, the fellow eye is observed. If the non-occluded eye displays the adjustment movement to regain fixation, the non-occluded eye is in manifest deviation. When the occluded eye is uncovered, movements of the uncovered eye are observed. While uncovering the eye, the examiner should monitor the possible adjustment movement to regain fixation and assert whether there is evidence of amblyopia. The longer the squinting eye keeps the fixation steady after uncovering the healthy eye, the greater the evidence that the visual acuity of the heterotropic eye is complementary to the fixating eye. If the

TABLE 1. Development of vision

Age (months)	Milestone
1	Sleeping Transient heterophoria Gaze directed downwards Monocular fixation Visual communication by staring at faces Horizontal tracking up to 90°
2	Staring, rare blinking Establishing saccades Vertical tracking Binocular fixation: establishing eye-to-eye contact with another person
3-4	Responsive smiling Observing one's own hands - brings the hands closer to the eyes Tracking along the arc of 180° Convergence Visually guided reaching for the object (good postural tone and reflexes required) Accommodation
5	Stereopsis Recognizing known and unknown faces Reaching for objects with certainty
6	Visual attention for near and distance Recognizing favorite people, objects, toys Pushing objects aside as a sign of rejection Observing the environment Playing "hide and seek" Complete conjugated eye movements fully established
24	Letter matching test possible in 2.5 years of age in 80% of cases

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uncovered eye instantly takes over fixation, a strong dominance is present (2). In other words, the longer the squinting eye is in the fixation position after uncovering the healthy eye, the less likely it is to have developed amblyopia (2, 33). A child usually rejects and tries to avoid the occlusion of the better eye and can be indifferent when occluding the amblyopic eye (34).

CONCLUSION

It is mandatory to note the earliest possible obstruction of the visual axis. Prompt referral to a paediatric ophthalmologist is of the utmost importance for improving the visual potential of the affected eye, and in cases of retinoblastoma, it can be a life-saving intervention. Key tests for visual assessment in infants are the red reflex test, fix and follow and the cover-uncover test. The described tests are simple and swift procedures and, as such, are important tools for the paediatrician. In cases of visual deprivation, immediate referral to a paediatric ophthalmologist is necessary.

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SUMMARY

Pregled vida u primarnoj pedijatrijskoj zdravstvenoj zaštiti

Dunja Bajtl, Mirjana Bjeloš, Mladen Bušić, Ana Ćurić, Vladimir Bajtl, Josip Barać

Uvod: Procjena vidne funkcije često predstavlja izazov za pedijatra u primarnoj zdravstvenoj zaštiti. Međutim, zahvaljujući jednostavnosti osnovnih metoda procjene vida, pedijatar može adekvatno evaluirati vidne funkcije u djece. Test crvenog refleksa kao vitalni dio svakog preventivnog pregleda djeteta u pedijatrijskoj ordinaciji primarne zdravstvene zaštite je najvažniji alat za otkrivanje opstrukcije vidne osi koja je uzrok nastanka deprivacijske slabovidnosti. Vidna os može biti opstruirana zbog različitih bolesti koje ugrožavaju vid ili život.

Cilj: Predstaviti metode procjene vidne oštirine dostupne pedijatru u primarnoj zdravstvenoj zaštiti.

Izvori podataka: PubMed, Google Scholar, Scopus, smjernice za preventivne preglede vida dostupne na internetu.

Zaključak: U slučajevima sumnje na opstrukciju vidne osi, urgentno upućivanje na pregled dječjem oftalmologu je obavezno.

Ključne riječi: DIJETE; TESTOVI VIDA; VIDNE FUNKCIJE; OFTALMOSKOP; AMBLIOPIJA