Chapter 6 Pupil Examination in Children

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ABSTRACT

Pupillary evaluation is a crucial part of every pediatric eye exam. It provides information about the structure of the eye as well as the afferent and efferent function of the eyes. It remains a quick and efficient test in the ocular evaluation. This chapter reviews the relevant neuroanatomy and the different components of the pupillary exam. The light-reflex and swinging flashlight tests are reviewed, as well as how a reverse afferent pupillary defect is detected. Clinical examples and pearls are provided to help the reader better understand the different aspects of the exam.

INTRODUCTION

Pupil testing is a crucial component of every pediatric eye examination. The shape and movements of the pupils provide an enormous amount of diagnostic information. Unequal pupil size (anisocoria) or a relative afferent pupillary defect (RAPD) may indicate a serious intracranial pathology or a benign physiological condition. An RAPD may be produced by retinal disorders (including but not limited to retinal detachment, and retinal vascular disease such as central retinal artery occlusion), unilateral or asymmetric optic nerve pathology, as well as chiasmal compression and optic tract lesions due to an uneven number of optic fibers undergoing hemidecussation at the chiasm (Balcer, 2019). An RAPD will NOT be produced by refractive error, media opacities including cataracts, lesions posterior to the lateral geniculate body and non-physiologic visual loss.

BACKGROUND

Relevant Neuroanatomy

Pupillary responses depend on the afferent and efferent neurologic pathways of the eyes, which will be reviewed in detail in this chapter.

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After light enters the eye, passing through cornea, anterior chamber, and lens it will eventually reach the retina, where it will stimulate the photoreceptors which will convert light stimuli to neuronal impulses. The afferent path originates at the level of the retina and sends neuronal impulses to various central nervous system structures, including the primary visual cortex for further processing and image recognition and the pretectal nuclei of the brainstem for control of pupillary constriction via the visceral efferent fibers of the oculomotor nerve (Agur and Dalley, 2019).

The pupillary-light reflex arc is polysynaptic and its afferent path is cranial nerve II and the efferent path is cranial nerve III (Figure 1). The afferent pathway starts at the level of the retina, ganglion cells project to the pretectal nuclei bilaterally in the midbrain. As nerve fibers pass through the chiasm hemidecussation of the nasal fibers occurs. The ganglion cell nerve fibers of the pupillary pathway exit the optic tract just anterior to the lateral geniculate nucleus at the brachium of the superior colliculus. The pretectal nuclei project crossed fibers via the posterior commissure, and uncrossed fibers to the Edinger-Westphal nucleus of cranial nerve III. The efferent path of the pupillary light reflex is the parasympathetic pathway which will be described in the following paragraphs.

The iris has both sympathetic and parasympathetic innervation. The size of the pupil depends on the balance between the sympathetic innervation to the dilator muscle of the iris and the parasympathetic innervation to the sphincter muscles of the iris.

Sympathetic innervation starts at the hypothalamus (McDougal and Gamlin, 2015). First-order neurons descend through the brain stem to the spinal cord in the intermediolateral column where they synapse in the ciliospinal nucleus of Budge-Waller, between the 7th cervical and the 2nd thoracic vertebrae. From there, the postsynaptic second-order fibers exit the spinal cord and ascend to terminate at the superior cervical ganglion at the level of the angle of the jaw and the carotid artery bifurcation. From the superior cervical ganglion, the postganglionic third-order fibers surround the internal carotid artery and enter the cavernous sinus. They briefly join cranial nerve VI, then join the nasociliary branch of the ophthalmic division of cranial nerve V, passing through the ciliary ganglion (without synapsing at the ciliary ganglion), before finally reaching the iris dilator muscle via the long ciliary nerves.

Parasympathetic innervation of the iris sphincter muscles originates at the Edinger-Westphal nucleus (McDougal and Gamlin, 2015). The preganglionic parasympathetic fibers travel within cranial nerve III, after the cranial nerve III bifurcation in the cavernous sinus the parasympathetic fibers travel in the inferior division of cranial nerve III until they reach and synapse at the ciliary ganglion. From there, the postsynaptic fibers will reach the iris sphincter via the short posterior ciliary nerves.

Clinically in the absence of ocular pathology, stimulation of one eye with light results in bilateral pupillary constriction. This occurs because of the double decussation of the fibers involved in the light reflex pathway, first at the chiasm and second at the posterior commissure where the pretectal nuclei project crossed fibers to the Edinger Westphal nucleus.

In addition to the pupillary-light reflex pathway, there is the near-reflex pathway which is responsible for pupillary constriction during accommodation (Kardon, 2008). Even though the neuroanatomic pathway responsible for the near-reflex is not as well defined, its efferent path is also through cranial nerve III with fibers originating in the rostral Edinger-Westphal nucleus.

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