HOSPITAL PHYSICIAN®

NEUROLOGY BOARD REVIEW MANUAL

Neuro-ophthalmology: Disorders of the Efferent Visual Pathway

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Table of Contents

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STATEMENT OF EDITORIAL PURPOSE

The *Hospital Physician Neurology Board Review Manual* is a peer-reviewed study guide for residents and practicing physicians preparing for board examinations in neurology. Each manual reviews a topic essential to the current practice of neurology.

NOTE FROM THE PUBLISHER:
This publication has been developed without involvement of or review by the American Board of Psychiatry and Neurology.

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next page

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INTRODUCTION

This manual is the second half of a 2-part review of neuro-ophthalmology. In the previous manual, diseases of the afferent visual pathway were considered. The afferent visual system includes the retina, optic nerve, optic tract, optic chiasm, and retrochiasmal pathways, including the optic radiations and the cortical/higher cognitive areas of visual representation. In this manual, the discussion focuses on diseases of the efferent visual system. The efferent component of the visual system includes the pupil and the associated autonomic pathways, neural mechanisms of ocular motility, and the cranial nerves involved in eyelid function. As in the previous manual, clinical cases are used to illustrate essential concepts guiding the physician to the location of pathology as well as the underlying process of disease.

DISEASE OF THE PUPIL

The diagnosis of a visual disturbance hinges on correctly identifying the site of pathology. Before considering specific disorders of the pupil, it is useful to begin with a brief review of the neuroanatomic pathways governing pupillary function.

CONTROL OF PUPILLARY FUNCTION

Pupillary size is under autonomic control. The parasympathetic pathway begins with light-induced activity in the retina, where signals are sent to the optic nerve and optic tract and finally to the pretectal nucleus of the midbrain and the Edinger-Westphal nucleus (via interneurons and the posterior commissure). Of note, stimulation of the Edinger-Westphal nucleus results in stimulation of the contralateral nucleus, thus producing a consensual response. Each Edinger-Westphal nucleus in turn transmits parasympathetic signals via the parasympathetic division of the ipsilateral oculomotor nerve (CN III). These parasympathetic fibers are found on the inferior portion of the oculomotor nerve. The axons synapse (activating nicotinic acetylcholine receptors) at the ciliary ganglion, which then sends short ciliary nerves (between the sclera and choroid) to the iris sphincter and ciliary muscles (activating muscarinic acetylcholine receptors). This results in pupillary constriction and lens accommodation, respectively.

The sympathetic pathway primarily begins in the insular cortex, which sends fibers to neurons in the hypothalamus. These hypothalamic cells are often considered the *central*, or *first-order, neurons* of the sympathetic pathway. From here, fibers are sent through the midbrain, pons, and lateral medulla to the intermediolateral neurons of the spinal cord (from the level of C8 to T2). The intermediolateral cells represent the *second-order neurons*, which in turn send projections exiting the cord at T1 to enter the sympathetic chain (via a white communicating ramus). The axons travel over the apex of the lung and continue through the inferior cervical ganglion to synapse on the superior cervical ganglion (activating nicotinic acetylcholine receptors). Cells in this latter ganglion represent the *third-order neurons*, which then send fibers up the internal carotid artery to the cavernous sinus. Here, the sympathetic fibers leave the carotid artery and travel first with the abducens nerve (CN VI) and then with the nasociliary nerve (which is a branch of the first division [V1] of the trigeminal nerve [CN V]) through the superior orbital fissure into the orbit. These axons reach (but do not synapse on) the ciliary ganglion in the orbit. At this point, sympathetic fibers from the ciliary ganglion (called the long ciliary nerve) finally reach the iris dilator muscles and activate noradrenergic receptors to cause pupillary dilation. These fibers also travel to the Müller's muscles of the upper and lower eyelid, assisting in widening the palpebral fissure (ie, keeping the eye open).

THE LARGE PUPIL

Case 1 Presentation

A previously healthy 34-year-old woman presents to the emergency department (ED) with a 3-hour history of gradual-onset blurry vision of the right eye, which began while she was reading. The patient says she noticed that her right pupil is larger