

HOSPITAL PHYSICIAN®

NEUROLOGY BOARD REVIEW MANUAL

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.

PUBLISHING STAFF

PRESIDENT, GROUP PUBLISHER

Bruce M. White

EDITORIAL DIRECTOR

Debra Dreger

ASSOCIATE EDITOR

Rita E. Gould

EDITORIAL ASSISTANT

Farrowh Charles

EXECUTIVE VICE PRESIDENT

Barbara T. White

EXECUTIVE DIRECTOR OF OPERATIONS

Jean M. Gaul

PRODUCTION DIRECTOR

Suzanne S. Banish

PRODUCTION ASSOCIATE

Kathryn K. Johnson

ADVERTISING/PROJECT DIRECTOR

Patricia Payne Castle

SALES & MARKETING MANAGER

Deborah D. Chavis

NOTE FROM THE PUBLISHER:

This publication has been developed without involvement of or review by the American Board of Psychiatry and Neurology.



Endorsed by the
Association for Hospital
Medical Education

Neuro-ophthalmology: Disorders of the Afferent Visual Pathway

Editor:

Alireza Atri, MD, PhD

Instructor in Neurology, Harvard Medical School; Assistant in Neurology, Memory Disorders Unit, Massachusetts General Hospital, Boston, MA

Associate Editor:

Tracey A. Milligan, MD

Instructor in Neurology, Harvard Medical School; Associate Neurologist, Brigham and Women's and Faulkner Hospitals, Boston, MA

Contributors:

Adam B. Cohen, MD

Chief Resident in Neurology, Harvard Medical School, Departments of Neurology, Massachusetts General and Brigham and Women's Hospitals, Boston, MA

Misha L. Pless, MD

Associate Professor of Neurology, Harvard Medical School; Director, Division of General Neurology, Department of Neurology, Massachusetts General Hospital, Boston, MA

Table of Contents

Introduction	2
Disease of the Retina	2
Disease of the Optic Nerve	4
Disease of the Optic Chiasm	6
Disease of the Retrochiasm/Cortical Pathways	10
Disease of Cerebrospinal Fluid Flow Dynamics	12
Summary	14
References	14

Cover Illustration by Kathryn K. Johnson

Copyright 2007, Turner White Communications, Inc., Strafford Avenue, Suite 220, Wayne, PA 19087-3391, www.turner-white.com. All rights reserved. No part of this publication may be reproduced, stored in a retrieval system, or transmitted in any form or by any means, mechanical, electronic, photocopying, recording, or otherwise, without the prior written permission of Turner White Communications. The preparation and distribution of this publication are supported by sponsorship subject to written agreements that stipulate and ensure the editorial independence of Turner White Communications. Turner White Communications retains full control over the design and production of all published materials, including selection of topics and preparation of editorial content. The authors are solely responsible for substantive content. Statements expressed reflect the views of the authors and not necessarily the opinions or policies of Turner White Communications. Turner White Communications accepts no responsibility for statements made by authors and will not be liable for any errors of omission or inaccuracies. Information contained within this publication should not be used as a substitute for clinical judgment.

Neuro-ophthalmology: Disorders of the Afferent Visual Pathway

Adam B. Cohen, MD, and Misha L. Pless, MD


INTRODUCTION

The visual system is a fundamental nervous system component. Neuro-ophthalmologic problems present special challenges in regard to the physician's knowledge of neuroanatomy and clinical examination skills. Viewing the fundus offers real-time inspection of the nervous system and its pathology. This view reveals a vast arrangement of neural structures dedicated to capturing, processing, and scrutinizing light. Given the evolutionary importance of vision, visual pathways occupy a great extent of the neurologic landscape. Thus, a keen understanding of vision is crucial for appreciating many, if not most, neurologic illnesses.

This manual, the first part of a 2-part review of neuro-ophthalmology, focuses on disorders of the afferent visual pathway and uses clinical cases to illustrate essential concepts guiding the physician to the location of pathology as well as the underlying process of disease. The afferent visual system includes the retina, optic nerve, optic tract, optic chiasm, and retrochiasm pathways, including the optic radiations and the cortical/higher cognitive areas of visual representation. The efferent visual system, which will be the focus of the next manual, encompasses the pupil as well as the mechanism of ocular motility and cranial nerves involved in eyelid function.

DISEASE OF THE RETINA

CASE 1 PRESENTATION

 A 66-year-old woman with a history of hypertension and lower extremity deep vein thrombosis (DVT) presents to the emergency department (ED) after a 5-minute episode of vision loss in the right eye. The patient had been well until earlier that day, when she developed sudden-onset loss of vision from the right eye beginning in the upper visual field. She covered her right eye and observed that vision from

her left eye was normal. The patient had no eye pain, change in cognition or language, weakness, or other difficulties. Spontaneous and rapid recovery of vision ensued, after which she noticed no visual or neurologic deficits.

- **What is the most likely cause of this patient's symptoms? What disorders are in the differential diagnosis?**

DIFFERENTIAL DIAGNOSIS OF TRANSIENT MONOCULAR VISUAL LOSS

In order to establish diagnostic possibilities in this patient, the evaluation begins with localizing the site of pathology. The patient's visual symptoms are consistent with transient monocular visual loss (TMVL), which may reflect pathology in various sites including the retina, optic nerve, and even the more posterior visual structures including the cortex. Given the brevity and acuity of her symptoms and the altitudinal pattern of visual loss, the most likely site of disease is the retinal arterial vessels. In this setting, the probable source of arterial disruption is atheroembolism from a stenotic ipsilateral internal carotid artery (ICA).

Vascular Causes

TMVL is most often considered a disease of the retina, and more specifically the retinal vasculature (eg, the ipsilateral ICA).¹⁻⁵ A severe stenosis of the ipsilateral ICA is made more likely when the patient reports rapid onset of symptoms and TMVL lasting 1 to 10 minutes.⁵ Visual loss respecting either the horizontal or vertical meridian commonly occurs in this setting and also points to disease of the ipsilateral ICA,⁴ although alternative sources of embolization are possible. Patients may perceive a curtain being pulled over the eye. Despite a lack of persistent symptoms, retinal emboli may be seen on fundoscopic examination (**Figure 1**; see page 8).

Additional important causes of TMVL via arterial disease of the retina or optic nerve include giant cell arteritis (GCA),^{6,7} other ischemic optic neuropathies,⁸ retinal