

HOSPITAL PHYSICIAN®

PULMONARY DISEASE BOARD REVIEW MANUAL

PUBLISHING STAFF

PRESIDENT, PUBLISHER
Bruce M. White

EXECUTIVE EDITOR
Debra Dreger

SENIOR EDITOR
Miranda J. Hughes, PhD

EDITOR
Becky Krumm

ASSISTANT EDITOR
Kathryn Charkatz

EDITORIAL ASSISTANT
Barclay Cunningham

SPECIAL PROGRAMS DIRECTOR
Barbara T. White, MBA

PRODUCTION MANAGER
Suzanne S. Banish

PRODUCTION ASSOCIATE
Vanessa Ray

ADVERTISING/PROJECT COORDINATOR
Patricia Payne Castle

NOTE FROM THE PUBLISHER:

This publication has been developed without involvement of or review by the American Board of Internal Medicine.



**Endorsed by the
Association for Hospital
Medical Education**

The Association for Hospital Medical Education endorses HOSPITAL PHYSICIAN for the purpose of presenting the latest developments in medical education as they affect residency programs and clinical hospital practice.

Alveolar Hemorrhage Syndromes

Series Editor:

Robert A. Balk, MD, FACP, FCCP, FCCM

Professor of Medicine, Director of Pulmonary and Critical Care Medicine, Department of Medicine, Rush Medical College and Rush-Presbyterian-St. Luke's Medical Center, Chicago, IL

Contributing Author:

David P. Gurka, MD, PhD, FCCP

Assistant Professor of Medicine, Section of Pulmonary and Critical Care Medicine, Department of Medicine, Rush Medical College and Rush-Presbyterian-St. Luke's Medical Center, Chicago, IL

Table of Contents

Introduction	2
Case Presentations	3
Clinical Features and Initial Evaluation	4
Diagnosis	6
Alveolar Hemorrhage Syndromes	7
Conclusion	10
Summary Points	10
Board Review Questions and Answers	10, 11
References	12
Suggested Readings	12

Cover Illustration by Dean Vigyikan

Copyright 1998, Turner White Communications, Inc., 125 Strafford Avenue, Suite 220, Wayne, PA 19087-3391. 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, Inc. The editors are solely responsible for selecting content. Although the editors take great care to ensure accuracy, Turner White Communications, Inc., will not be liable for any errors of omission or inaccuracies in this publication. Opinions expressed are those of the authors and do not necessarily reflect those of Turner White Communications, Inc.

HOSPITAL PHYSICIAN®

PULMONARY DISEASE BOARD REVIEW MANUAL

Alveolar Hemorrhage Syndromes

Series Editor:

Robert A. Balk, MD, FACP, FCCP, FCCM
Professor of Medicine
Director of Pulmonary and Critical Care Medicine
Department of Medicine
Rush Medical College and
Rush-Presbyterian-St. Luke's Medical Center
Chicago, IL

Contributing Author:

David P. Gurka, MD, PhD, FCCP
Assistant Professor of Medicine
Section of Pulmonary and Critical Care Medicine
Department of Medicine
Rush Medical College and
Rush-Presbyterian-St. Luke's Medical Center
Chicago, IL

I. INTRODUCTION

Alveolar hemorrhage is the occurrence of blood arising from and pooling in pulmonary acinar sacs. This relatively uncommon finding is often associated with extrapulmonary and/or laboratory abnormalities that characterize a diverse group of disorders known as alveolar (or pulmonary) hemorrhage syndromes. Alveolar hemorrhage syndromes are frequently immune-mediated but may also involve drug or toxin exposure. Alveolar hemorrhage is often associated with renal disease.

There is no universally accepted classification scheme for alveolar hemorrhage syndromes, although in general they are grouped by extrapulmonary clinical manifestations or by the histologic presence or absence of capillaritis (**Table 1**). Pulmonary capillaritis was first described by Spencer in 1957, in association with polyarteritis nodosa. The concept was refined by Mark and Ramirez in 1985 in their report on patients with systemic vasculitides.

Pulmonary capillaritis is characterized by infiltration of the alveolar interstitium by neutrophils, which leads to fibrinoid necrosis of the capillary walls with leakage of erythrocytes and neutrophils into the alveolar septa and spaces. Fibrin thrombi can be found occluding the septal capillaries as well as lining the alveolar sacs. Erythrocytes are present in the interstitium and alveolar spaces in the

presence of acute hemorrhage, and hemosiderin-laden macrophages and free hemosiderin are markers of recent or chronic hemorrhage. Inflammation of the pulmonary vasculature is usually indicative of a systemic vasculitis.

Although any form of bleeding diathesis or other condition that increases pulmonary capillary pressure (ie, mitral stenosis, pulmonary veno-occlusive disease, pulmonary capillary hemangiomatosis) can manifest as alveolar hemorrhage, this review will be restricted to those entities, with as well as without capillary inflammation, in which the alveolar epithelium or capillary endothelium is directly involved. Because the differential diagnosis of alveolar hemorrhage is broad (**Table 1**), only the more common of these relatively rare syndromes will be discussed.

The relative frequencies of the causes of alveolar hemorrhage are not well delineated. In one 6-year series of patients from the University of Minnesota, the most common cause of alveolar hemorrhage was anti-glomerular basement membrane (anti-GBM) antibody disease, followed by the systemic vasculitides (many types), pauci-immune glomerulonephritis, and connective tissue diseases (eg, systemic lupus erythematosus).¹ Another review, however, found Wegener's disease and other vasculitides to be nearly three times as prevalent as anti-GBM antibody disease as a cause of alveolar hemorrhage.²