

Approach to Internal Medicine

A Resource Book for Clinical
Practice

David Hui · Alexander A. Leung ·
Christopher Ma *Editors*

Fifth Edition



Springer

Editors

David Hui
The University of Texas MD
Anderson Cancer Center
Houston, TX
USA

Alexander A. Leung
University of Calgary
Calgary, AB
Canada

Christopher Ma
University of Calgary
Calgary, AB
Canada

ISBN 978-3-030-72979-0 ISBN 978-3-030-72980-6 (eBook)
<https://doi.org/10.1007/978-3-030-72980-6>

© The Editor(s) and The Author(s), under exclusive license to Springer Nature
Switzerland AG 2022

This work is subject to copyright. All rights are solely and exclusively licensed by the Publisher, whether the whole or part of the material is concerned, specifically the rights of translation, reprinting, reuse of illustrations, recitation, broadcasting, reproduction on microfilms or in any other physical way, and transmission or information storage and retrieval, electronic adaptation, computer software, or by similar or dissimilar methodology now known or hereafter developed.

The use of general descriptive names, registered names, trademarks, service marks, etc. in this publication does not imply, even in the absence of a specific statement, that such names are exempt from the relevant protective laws and regulations and therefore free for general use.

The publisher, the authors and the editors are safe to assume that the advice and information in this book are believed to be true and accurate at the date of publication. Neither the publisher nor the authors or the editors give a warranty, expressed or implied, with respect to the material contained herein or for any errors or omissions that may have been made. The publisher remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

This Springer imprint is published by the registered company Springer Nature Switzerland AG
The registered company address is: Gewerbestrasse 11, 6330 Cham, Switzerland

Pulmonary Hypertension

Hirani et al. *Can J Cardiol* 2020;36(7)

WHO CLASSIFICATION OF PULMONARY HYPERTENSION

GROUP I. PULMONARY ARTERIAL HYPERTENSION

- **IDIOPATHIC**—primary
- **FAMILIAL DISORDERS**
- **DRUG AND TOXIN INDUCED**
- **PAH ASSOCIATED WITH SPECIFIC DISORDERS**—connective tissue disease, HIV, portal hypertension, congenital heart disease, schistosomiasis
- **PAH LONG TERM RESPONDERS TO CALCIUM CHANNEL BLOCKERS**
- **PAH WITH SIGNIFICANT VENOUS OR CAPILLARY INVOLVEMENT**—pulmonary veno-occlusive disease, pulmonary–capillary hemangiomatosis
- **PERSISTENT PULMONARY HYPERTENSION OF NEWBORN**

GROUP II. PULMONARY VENOUS HYPERTENSION DUE TO LEFT HEART DISEASE

heart disease with preserved LVEF, heart disease with reduced LVEF, valvular heart disease, cardiovascular conditions leading to postcapillary PH

GROUP III. PULMONARY HYPERTENSION DUE TO LUNG DISEASE AND/OR HYPOXEMIA

obstructive lung disease, restrictive lung disease, mixed restrictive/obstructive disease, including obstructive sleep apnea and obesity hypoventilation syndrome, developmental lung disease

GROUP IV. PULMONARY HYPERTENSION DUE TO PULMONARY ARTERY OBSTRUCTIONS

chronic thromboembolic, other pulmonary artery obstructions (i.e. tumor, parasites, foreign material)

GROUP V. PULMONARY HYPERTENSION WITH UNCLEAR AND/OR MULTIFACTORIAL MECHANISMS

hematological (pulmonary Langerhans cell histiocytosis, lymphangiomatosis), systemic and metabolic disorders (sarcoidosis),

WHO CLASSIFICATION OF PULMONARY HYPERTENSION (CONT'D)

complex congenital heart disease, others (compression of pulmonary vessels by tumor, fibrosing mediastinitis)

PATHOPHYSIOLOGY

DEFINITION OF PULMONARY HYPERTENSION—mean pulmonary arterial pressure (PAP) >25 mmHg at rest or mean PAP >30 mmHg with exercise measured with right heart catheterization

CLINICAL FEATURES

HISTORY—unexplained dyspnea on exertion, cough, chest pain, hemoptysis, dizziness, syncope, hoarseness, past medical history (cardiac and respiratory diseases, thromboembolic diseases, HIV, cirrhosis, autoimmune and rheumatologic disorders), medications (amphetamine, diet pill such as dexfenfluramine)

PHYSICAL—vitals (tachypnea, tachycardia, atrial fibrillation, hypoxemia), peripheral cyanosis, small pulse volume, elevated JVP (prominent a wave or absent if atrial fibrillation, large v wave), right ventricular heave, loud or palpable P2, right-sided S4, tricuspid regurgitation murmur, Graham-Steell murmur (high-pitched, decrescendo diastolic rumble over LUSB), crackles, congestive liver, ascites, ankle edema

INVESTIGATIONS

BASIC

- **LABS**—CBC, lytes, urea, Cr, AST, ALT, ALP, bilirubin, INR, albumin, ANA, RF, anti-CCP, anti-Scl-70, anticentromere antibody, ESR, HIV serology, TSH
- **IMAGING**—CXR, CT chest, V/Q scan or CT chest PE protocol, echocardiogram

INVESTIGATIONS (CONT'D)

- **ECG**
- **OVERNIGHT POLYSOMNOGRAPHY**—if suspect OSA
- **ABG**
- **PFT**

SPECIAL

- **RIGHT HEART CATHETERIZATION WITH VASOREACTIVITY TESTING**

MANAGEMENT

SYMPTOM CONTROL—**diuretics**, **O₂**, **anticoagulation**, **calcium channel blockers** if positive vasoreactivity test (in high doses), **endothelin receptor antagonists** (bosentan,

MANAGEMENT (CONT'D)

ambrisentan), **phosphodiesterase type-5 inhibitors** (sildenafil), prostacyclin analogues (epoprostenol, iloprost, selexipag), soluble guanylate cyclase stimulators (riociguat)

TREAT UNDERLYING CAUSE**ATRIAL SEPTOSTOMY****LUNG TRANSPLANT****REFERRAL TO A SPECIALIZED PULMONARY HYPERTENSION CLINIC****SPECIFIC ENTITIES**

EISENMENGER SYNDROME—left-to-right shunt leading to pulmonary hypertension and eventually right-to-left shunt