

# Contents

## Part I Introduction

- 1 Historical Perspective on the Classification and Nomenclature of Pulmonary Hypertension** ..... 3  
Myung H. Park
- 2 The Defining Characteristics of Pulmonary Arterial Hypertension** ..... 17  
Rahul Kumar, Jeffrey C. Robinson, and Rubin M. Tuder
- 3 Pulmonary Hypertension in Patients Without Pulmonary Arterial Hypertension** ..... 29  
Jessica Huston, Nathan D. Hatton, and John J. Ryan
- 4 Epidemiology of Pulmonary Hypertension: From Quaternary Referral Centre to the Community** ..... 63  
Gaurav Choudhary and Corey E. Ventetuolo

## Part II Basic Mechanism of Disease

- 5 The Effects of Chronic Hypoxia on Inflammation and Pulmonary Vascular Function** ..... 83  
Kurt R. Stenmark, Steven C. Pugliese, Jens Poth, Maria G. Frid, Evgenia Gerasimovskaya, Eva Nozik-Grayck, and Karim C. El Kasmi
- 6 Genetics of Pulmonary Vascular Disease** ..... 105  
Lijiang Ma and Wendy K. Chung
- 7 Novel Mechanisms of Disease: Network Biology and MicroRNA Signaling in Pulmonary Hypertension** ..... 123  
Wassim H. Fares, Kusum V. Pandit, and Naftali Kaminski
- 8 Pulmonary Hypertension as a Metabolic Disease** ..... 135  
Joshua P. Fessel and William M. Oldham
- 9 Renin-Angiotensin-Aldosterone and Other Neurohumoral Factors in the Pathogenesis of Pulmonary Hypertension** ..... 147  
Bradley A. Maron, Thomas E. Stephens, and Jane A. Leopold

## Part III Translational and Clinical Principles of Pulmonary Vascular Disease and Right Ventricular Dysfunction

- 10 Animal Models of Pulmonary Hypertension** ..... 161  
Jose Gomez-Arroyo, Ivana Nikolic, and Paul B. Yu

<b>11</b>	<b>The Cardiopulmonary Hemodynamic Evaluation of Pulmonary Hypertension</b> .....	173
	Yonatan Buber and Alexander R. Opatowsky	
<b>12</b>	<b>Advanced Imaging in Pulmonary Hypertension</b> .....	199
	Geeshath Jayasekera and Andrew J. Peacock	
<b>13</b>	<b>Assessing Disease State in the Pulmonary Vasculature in Clinical Practice and Research</b> .....	219
	Manyoo Agarwal and Aaron B. Waxman	
<b>14</b>	<b>Biomarkers and Other Methods for Assessing Patient Progress</b> .....	231
	Nadine Al-Naamani and Ioana R. Preston	
<b>15</b>	<b>Pulmonary Circulatory – Right Ventricular Uncoupling: New Insights Into Pulmonary Hypertension Pathophysiology</b> .....	241
	David Boulate, Olaf Mercier, Julien Guihaire, Elie Fadel, Robert Naeije, Francois Haddad, and Franz Rischard	
 <b>Part IV Clinical Evaluation and Management of Pulmonary Hypertension</b>		
<b>16</b>	<b>Contemporary Pharmacotherapies Involving Nitric Oxide, Prostacyclin, and Endothelin Receptor Signaling Pathways</b> .....	257
	Nicholas M. Furiasse and Jonathan D. Rich	
<b>17</b>	<b>Determining the Optimal Approach to Initiating Oral, Inhaled, and Intravenous Therapies in Clinical Practice: Sequential Goal-Directed Therapy Is Best</b> .....	271
	Brian B. Graham	
<b>18</b>	<b>Determining the Optimal Approach to Initiating Oral, Inhaled, and Intravenous Therapies in Clinical Practice: Maximal Upfront Therapy Is Best</b> .....	277
	Victor F. Tapson	
<b>19</b>	<b>Surgery, Devices, Transplantation and Other Interventional Options for the Treatment of Advanced Pulmonary Hypertension</b> .....	283
	Christian Bermudez and Balakrishnan Mahesh	
<b>20</b>	<b>Patient Registries in Pulmonary Arterial Hypertension: the Role of Survival Equations and Risk Calculators</b> .....	307
	Colleen A. McEvoy, Mayank Sardana, Matthew Moll, Harrison W. Farber, and Murali M. Chakinala	
<b>21</b>	<b>Support Care for the Pulmonary Hypertension Patient</b> .....	327
	Abby Poms, Mary Bartlett, Traci Houston, Martha Kingman, and Glenna L. Traiger	
<b>22</b>	<b>Special Considerations for the Pulmonary Hypertension Patient</b> .....	345
	Kristina T. Kudelko, Roham T. Zamanian, and Vinicio A. De Jesus Perez	
<b>23</b>	<b>The Future of Pulmonary Hypertension</b> .....	359
	Robert P. Frantz	
<b>Index</b> .....		369