

Case Report

Growing up with Idiopathic Pulmonary Arterial Hypertension: An Arduous Journey

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Abstract: Idiopathic pulmonary arterial hypertension (IPAH) is an uncommon and severe disease. We report the case of a 7-year-old boy investigated for cardiac murmur and exercise intolerance. Pulmonary hypertension (PH) was suspected at clinical examination and confirmed by echocardiography and cardiac catheterization. This case of pulmonary hypertension was classified as idiopathic given the negative etiological investigation. Vasoactive testing with oxygen and nitric oxide was negative. Therefore, treatment with sildenafil (1.4 mg/kg/d) and bosentan (3 mg/kg/d) was initiated. This allowed the stabilization of, but not a decrease in, pulmonary artery pressure for the next 5 years, during which the patient's quality of life was significantly reduced. At a later follow-up, the estimated pulmonary pressure was found to have increased and become supra-systemic, with a consequent deterioration in the child's condition. This led to the decision to enter him into a clinical trial that is still ongoing. Idiopathic pulmonary arterial hypertension is a severe disease that can present with non-specific symptoms, such as asthenia and exercise limitation, which are important not to trivialize. The disease is associated with significantly decreased quality of life in affected children and carries a high burden in terms of mortality and morbidity. The current knowledge about IPAH in children is reviewed, with a particular focus on the future prospects for its treatment and the related quality of life of patients.

Keywords: pulmonary arterial hypertension; idiopathic; children; vasoactivity; treatment; quality of life



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1. Introduction

Pulmonary hypertension is a rare but severe condition defined by a mean pulmonary arterial pressure (mPAP) > 20 mmHg at rest [1].

Depending on the etiology, five different groups of PH are described and listed in Table 1 [1,2]: pulmonary arterial hypertension (PAH) (group 1), PH associated with left heart disease (group 2), PH associated with lung disease and/or hypoxemia (group 3), PH associated with pulmonary artery obstruction (group 4), and PH with unclear and/or multifactorial mechanisms (group 5) [1–4].

PAH or pre-capillary pulmonary hypertension, which includes idiopathic pulmonary hypertension (IPAH), is defined as pulmonary hypertension with a pulmonary-artery-wedge pressure ≤ 15 mmHg with PVR ≥ 2 WU (2), excluding groups 4 and 5.

Idiopathic pulmonary arterial hypertension is a rare disease with an incidence ranging between 0.47 and 1–2 cases per million children, and with a prevalence of 2.1 to 4.4 cases per million children [2].

Although most children with PAH can now reach adulthood, thanks to recent therapeutic advances, the burden created by the disease in terms of morbidity and mortality remains high [5].

A Journey With Pulmonary Arterial Hypertension

Lung Recipients Around the World



A Journey With Pulmonary Arterial Hypertension:

Murray & Nadel's Textbook of Respiratory Medicine E-Book V. Courtney Broaddus, Joel D. Ernst, Talmadge E. King Jr, Stephen C. Lazarus, Kathleen F. Sarmiento, Lynn M. Schnapp, Renee D. Stapleton, Michael B. Gotway, 2021-05-28 Known for its clear readability thorough coverage and expert authorship Murray advanced ultrasound applications and procedures interventional pulmonology immunotherapy lung cancer targeted therapy outbreaks pandemics and bioterrorism point of care ultrasound use of high flow oxygen and more Includes extensively reorganized sections on basic science pleural disease and sleep with new chapters and approaches to the topics Features more than 1 450 anatomic algorithmic and radiologic images 400 are new including CT PET MR and HRCT plus extensive online only content 200 procedural and conceptual videos plus audio clips of lung sounds Brings you up to date with the latest respiratory drugs mechanisms of action indications precautions adverse effects and recommendations with increased emphasis on algorithms to illustrate decision making Enhanced eBook version included with purchase Your enhanced eBook allows you access to all of the text figures reporting templates and references from the book on a variety of devices Coronary Road: A Comprehensive Guide to the Molecular Basis of Cardiology Pasquale De Marco, 2025-04-13 Coronary Road A Comprehensive Guide to the Molecular Basis of Cardiology is an essential resource for cardiologists medical students and researchers seeking a deeper understanding of the molecular mechanisms underlying cardiovascular diseases Delving into the molecular architecture of the heart this book unveils the intricate interplay of molecular components that orchestrate the heart's function Readers will gain insights into the molecular pathways involved in cardiovascular diseases providing a solid foundation for understanding the pathophysiology of these conditions Coronary Road also explores the molecular pharmacology of cardiovascular drugs delving into the intricate mechanisms by which these medications exert their therapeutic effects The book provides insights into the molecular techniques employed in cardiology highlighting their role in diagnosis prognosis and treatment Furthermore the book explores the frontiers of molecular cardiology showcasing the potential of cutting edge technologies and emerging therapies to revolutionize the field From gene therapy to stem cell therapy readers will gain an understanding of the latest advancements poised to transform the landscape of cardiovascular care Written in an engaging and accessible style Coronary Road offers a unique perspective on the molecular underpinnings of cardiovascular disorders Its comprehensive coverage encompasses the molecular basis of atherosclerosis the genetic factors contributing to heart failure the molecular mechanisms of cardiac arrhythmias hypertension cardiomyopathies and congenital heart defects With its comprehensive coverage up to date information and engaging writing style Coronary Road is an indispensable guide for those seeking to expand their knowledge of molecular cardiology It is an invaluable resource for healthcare professionals researchers and students alike If you like this book write a review on google books Textbook of Pulmonary and Critical Care Medicine Vols 1 and 2 SK Jindal, PS Shankar, Suhail Raoof, Dheeraj Gupta, 2011-03-20 Book includes the basic

principles of Pulmonology as well as the recent advances in allied clinical sciences relevant to pulmonology Includes valuable inputs on tuberculosis other pulmonary infections environmental and occupational medicine sleep disorders and general systemic diseases affecting the respiratory system Although critical care is relevant for most of the medical and surgical specialties the pulmonologist have a more vested interest than other specialists Assisted respiration which forms the core of most critical care lies in the primary domain of pulmonologists

Harper's Practical Genetic Counselling, Eighth Edition Angus Clarke, 2016-06-15 Easy to use and useful when kept close at hand in the room where you work The book is a pleasure to read the style elegant and authoritative Lancet this book is a wonderful reference to enable primary physicians to be informed about their patients Annals of Internal Medicine Universally used across the world by genetic counsellors medical geneticists and clinicians alike Harper s Practical Genetic Counselling has established itself as the essential guide to counselling those at risk from inherited disorders Increasingly common disorders are known to have a genetic component and this book provides invaluable and up to date guidance through the profusion of new information in this area and the associated psychosocial and ethical considerations and concerns Within its established tried and trusted framework the book contains new chapters on laboratory methods new genetic sequencing techniques and the applications of genome wide SNP association studies genetic susceptibility cross cultural aspects and the genetic counselling process It has expand chapters on genetic screening and screening of newborn treatment techniques and rational approaches to treatment non Mendelian inheritance free fetal DNA in prenatal screening and diagnosis Key features Fully updated to provide the very latest information when in a busy consulting room or clinic Clear and authoritative advice applicable to everyday clinical practice Reflects the rapid development of knowledge in this area including the implications of the human genome project and related technology The eighth edition of this popular best selling text continues to be an essential source of reference for trainee and practitioner genetic counsellors medical geneticists and clinicians Also it provides valuable background for specialist nurses counsellors social scientists ethicists as well as genetics laboratory staff

Travel Medicine E-Book Jay S. Keystone, Phyllis E. Kozarsky, Bradley A. Connor, Hans D. Nothdurft, Marc Mendelson, Karin Leder, 2018-11-22 Includes new chapters to assist your care of specific populations such as those engaging in ecotourism or military travel as well as the VIP traveler A new chapter on pre travel considerations for non vaccine preventable travel infections has also been added Provides new information on new influenza and shingles vaccines microbiome and drug resistance Zika and the pregnant or breastfeeding traveler the Viagra effect and increase in STIs refugees and immigrants and much more Covers new methods of prevention of dengue virus Zika virus chikungunya virus Middle Eastern respiratory syndrome sleeping sickness and avian flu New illustrations and numerous new tables and boxes provide visual guidance and make reference quick and easy Helps you prepare for the travel medicine examination with convenient cross references to the ISTM body of knowledge in specific chapters and or passages in the book Keeps you updated on remote destinations and the unique perils they present

Mosby's 2018 Nursing Drug Reference - E-Book Linda Skidmore-Roth, 2017-04-25 Choose the drug handbook trusted for nearly 30 years From the leading name in nursing Mosby's 2018 Nursing Drug Reference is a full color portable nursing drug handbook that makes it easy to find the most vital information on the drugs that nurses administer most frequently More than 5 000 drugs are profiled including 20 30 new entries for drugs recently approved by the FDA No other drug guide places a higher emphasis on patient safety with Black Box Warnings for dangerous adverse reactions Safety Alerts for situations requiring special attention and a focus on both common and life threatening side effects Backed with practical online resources Mosby's Nursing Drug Reference proves there is a difference in drug guides It's Mosby's bestselling drug reference for nurses Black Box Warnings provide alerts to FDA warnings of dangerous or life threatening drug reactions Safety Alert icon highlights the most critical drug interactions and side effects Common and life threatening side effects are organized by body system showing signs to watch for during assessments Coverage of IV drug administration highlights dosage and IV administration instructions including safety considerations and Y site syringe and additive compatibilities Alphabetical organization by generic name provides quick and easy access to specific drugs and a full color design highlights important information Nursing Process steps are used as the framework for organizing all nursing care information Overview of drug categories explains the safe administration of common classes of drugs as well as their common side effects and interactions Cross references indicate drug information that may be found in the appendices Complete pharmacokinetic information includes the mechanism and absorption of the drug as well as its action duration and excretion NEW 20 30 added monographs provide up to date content on newly released drugs NEW BEERS indications included for selected monographs NEW Drugs with genetic indications identifiable by easy to spot icon

Mosby's 2020 Nursing Drug Reference E-Book Linda Skidmore-Roth, 2019-06-12 Put the most trusted name in nursing in your hands Mosby's 2020 Nursing Drug Reference is a full color portable nursing drug handbook that makes it easy to find the most vital information on the drugs that nurses administer most frequently More than 5 000 drugs are profiled in the text and on the free app including 20 30 new entries for drugs and drug therapies recently approved by the FDA Plus no other drug guide places a higher emphasis on patient safety with Black Box Warnings for dangerous adverse reactions Safety Alerts for situations requiring special attention and a focus on both common and life threatening side effects of 50 common drug classes This 33rd edition proves once again why Mosby's is the bestselling drug reference for nurses on the market Coverage of more than 5 000 generic and trade name drugs provides information for almost every drug a student is likely to encounter in clinicals Common and life threatening side effects are organized by body system showing signs to watch for during assessments IV drug administration coverage in bold highlight dosage and IV administration instructions including safety considerations and Y site syringe and additive compatibilities Safety Alert icon highlights the most critical drug interactions and side effects Nursing Process steps are used as the framework for organizing all nursing care information Complete pharmacokinetic information includes the mechanism

and absorption of the drug as well as its action duration and excretion Black Box Warnings provide alerts to FDA warnings of dangerous or life threatening drug reactions Logical alphabetical organization by generic name provides quick and easy access to specific drugs and a full color design highlights important information Cross references indicate drug information that may be found in the appendices NEW 20 30 added monographs provide up to date content on newly released FDA approved drugs and drug therapies NEW More than 20 new Evolve only drug monographs added to the existing Evolve only drug monographs NEW Numerous herbal drug monographs included on Evolve site

Congenital and Paediatric Acquired Heart Disease in Practice Yiu-fai Cheung,2023-09-11 This book provides concise practical and up to date information on congenital and acquired heart disease in children Thirty eight congenital cardiovascular malformations 9 acquired cardiovascular conditions physiology unique to congenital heart disease 10 common postoperative issues and topical issues including COVID 19 and the heart and lymphatic disorders and interventions in congenital heart patients are included Each chapter is in the reader friendly format covering aetiology pathophysiology clinical manifestations investigations and management approach of congenital and paediatric acquired heart disease by incorporation of coloured figures tables and boxes It will equip medical undergraduates and trainees in paediatric cardiology cardiac surgery cardiac intensive care and cardiac nursing staff with up to date knowledge and practical skills to manage and to care for children with congenital and acquired heart disease

The Right Heart - Pulmonary Circulation Unit, An Issue of Heart Failure Clinics Eduardo Bossone,Luna Gargani,2018-07-15 This issue of Heart Failure Clinics edited by Dr Eduardo Bossone will cover The Right Heart Pulmonary Circulation Unit Topics include Pathophysiology Increased Systemic versus Increased Pulmonary Pressures Pulmonary Arterial Hypertension Right Heart Pulmonary Circulation Unit in Connective Tissue Disease Right Heart Pulmonary Circulation Unit in Congenital Heart Diseases Pulmonary Hypertension and Heart Failure Right Heart Pulmonary Circulation Unit in Cardiomyopathies and Storage Diseases Pulmonary Hypertension Right Heart Pulmonary Circulation Unit at High Altitude Chronic Thromboembolic Pulmonary Hypertension Combining Invasive and Non Invasive Evaluation for the Diagnosis of Pulmonary Hypertension Imaging the Right Heart Pulmonary Circulation Unit The Role of Ultrasound Imaging the Right Heart Pulmonary Circulation Unit The Role of CT and MRI Biomarkers in Pulmonary Hypertension Pulmonary Hypertension Related to Diffuse Parenchymal Lung Disease Chronic Right Heart Failure Exercise Training and Rehabilitation in Pulmonary Hypertension and Right Heart Circulation Unit and Left Heart Valvular Diseases

Conn's Current Therapy 2020, E-Book Rick D. Kellerman,KUSM-W Medical Practice Association,2019-12-07 Designed to suit a wide range of healthcare providers including primary care subspecialties and allied health Conn's Current Therapy has been a trusted clinical resource for more than 70 years The 2020 edition continues this tradition of excellence with current evidence based treatment information presented in a concise yet in depth format More than 300 topics have been carefully reviewed and updated to bring you state of the art information even in the most rapidly changing areas of medicine Offers

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When Experiments Travel Adriana Petryna, 2024-11-01 The phenomenal growth of global pharmaceutical sales and the quest for innovation are driving an unprecedented search for human test subjects particularly in middle and low income countries Our hope for medical progress increasingly depends on the willingness of the world's poor to participate in clinical drug trials While these experiments often provide those in need with vital and previously unattainable medical resources the outsourcing and offshoring of trials also create new problems In this groundbreaking book anthropologist Adriana Petryna takes us deep into the clinical trials industry as it brings together players separated by vast economic and cultural differences Moving between corporate and scientific offices in the United States and research and public health sites in Poland and Brazil **When Experiments Travel** documents the complex ways that commercial medical science with all its benefits and risks is being integrated into local health systems and emerging drug markets Providing a unique perspective on globalized clinical trials **When Experiments Travel** raises central questions Are such trials exploitative or are they social goods How are experiments controlled and how is drug safety ensured And do these experiments help or harm public health in the countries where they are conducted Empirically rich and theoretically innovative the book shows that neither the language of coercion nor that of rational choice fully captures the range of situations and value systems at work in medical experiments today **When Experiments Travel** challenges conventional understandings of the ethics and politics of transnational science and changes the way we think about global medicine and the new infrastructures of our lives

Principles and Practice of Travel Medicine Jane N. Zuckerman, 2013-02-25 **Principles and Practice of Travel Medicine** This second edition of **Principles and Practice of Travel Medicine** has been extensively updated to provide a comprehensive description of travel medicine and is an invaluable reference resource to support the clinical practice of travel medicine This new edition covers the many recent advances in the field including the development of new and combined vaccines malaria prophylaxis emerging new infections new hazards resulting from travel to long haul destinations health tourism and population movements The chapter on vaccine

preventable diseases includes new developments in licensed vaccines as well as continent based recommendations for their administration There are chapters on the travel health management of high risk travellers including the diabetic traveller the immunocompromised those with cardiovascular renal neurological gastrointestinal malignant and other disorders psychological and psychiatric illnesses pregnant women children and the elderly With increasing numbers of ever more adventurous travellers there is discussion of travel medicine within extreme environments whilst the chapter on space tourism may well be considered the future in travel medicine Principles and Practice of Travel Medicine is an invaluable resource for health care professionals providing advice and clinical care to the traveller Titles of related interest Atlas of Human Infectious Diseases Heiman F L Wertheim Peter Horby John P Woodall 9781405184403 2012 Infectious Diseases A Geographic Guide Eskild Petersen Lin H Chen Patricia Schlagenhauf 9780470655290 2011 Tropical Diseases in Travelers Eli Schwartz 9781405184410 2009 For more information on all our resources in Infectious Diseases please visit www.wiley.com/go/infectiousdiseases

Travel Medicine Jay S. Keystone, David O Freedman, MD, Phyllis E. Kozarsky, MD, Bradley A. Connor, MD, Hans D. Nothdurft, MD, 2012-12-26 Travel Medicine 3rd Edition by Dr Jay S Keystone Dr Phyllis E Kozarsky Dr David O Freedman Dr Hans D Nothdurft and Dr Bradley A Connor prepares you and your patients for any travel related illness they may encounter Consult this one stop resource for best practices on everything from immunizations and pre travel advice to essential post travel screening From domestic cruises to far flung destinations this highly regarded guide offers a wealth of practical guidance on all aspects of travel medicine This is an excellent reference source that contains words of wisdom which covers an area of medicine which can sometimes get lost on the radar screen Reviewed by Dr Harry Brown on behalf of Glycosmedia Apr 2014 Benefit from the advice of international experts on the full range of travel related illnesses including cruise travel bird flu SARS traveler s diarrhea malaria environmental problems and much more Prepare for the travel medicine examination with convenient cross references for the ISTM body of knowledge to specific chapters and or passages in the book Search the complete text and download images at expertconsult.com Effectively protect your patients before they travel with new information on immunizations and emerging and re emerging disease strains including traveler s thrombosis Update your knowledge of remote destinations and the unique perils they present Stay abreast of best practices for key patient populations with new chapters on the migrant patient humanitarian aid workers medical tourism and mass gatherings as well as updated information on pediatric and adolescent patients

Taking Flight: Inspirational Stories of Lung Transplantation More Journeys Lung Recipients Around the World, 2011-05-06 I have the back cover designer working on the wording for the back cover I require no help [Conn's Current Therapy 2022 - E-Book](#) Rick D. Kellerman, David P. Rakel, KUSM-W Medical Practice Association, 2021-12-21 Trusted by clinicians for nearly 75 years Conn's Current Therapy presents today's evidence based information along with the personal experience and discernment of expert physicians The 2022 edition is an excellent resource for a wide range of healthcare providers including primary care

subspecialists and allied health providing current treatment information in a concise yet in depth format More than 300 topics have been carefully reviewed and updated to bring you state of the art content in even the most rapidly changing areas of medicine Offers personal approaches from recognized leaders in the field covering common complaints acute diseases and chronic illnesses along with the most current evidence based clinical management options Follows a consistent easy to use format throughout with diagnosis therapy drug protocols and treatment pearls presented in quick reference boxes and tables for point of care answers to common clinical questions Includes new and significantly revised chapters on COVID 19 and post COVID syndrome pyoderma gangrenosum mitochondrial disease gender affirming care stem cell therapy and artificial intelligence Incorporates more electronic links throughout the text that connect the reader to apps and clinical prediction tools that can easily be accessed in practice Features thoroughly reviewed and updated information from many new authors who offer a fresh perspective and their unique personal experience and judgment Provides current drug information thoroughly reviewed by PharmDs Features nearly 300 images including algorithms anatomical illustrations and photographs that provide useful information for diagnosis

Ward, Milledge and West's High Altitude Medicine and Physiology Andrew M Luks, Philip N Ainslie, Justin S Lawley, Robert C Roach, Tatum S Simonson, 2021-02-15 This pre eminent work has developed over six editions in response to man s attempts to climb higher and higher unaided and to spend more time at altitude for both work and recreation Building on this established reputation the new and highly experienced authors provide a fully revised and updated text that will help doctors continue to improve the health and safety of all people who visit live or work in the cold thin air of high mountains The sixth edition remains invaluable for any doctor accompanying an expedition or advising patients on a visit to altitude those specialising in illness and accidents in high places and for physicians and physiologists who study our dependence on oxygen and the adaptation of the body to altitude *Acute Respiratory Distress Syndrome* Sadis Matalon, Jacob Iasha Sznajder, 2012-12-06 Proceedings of a NATO ASI held in Corfu Greece June 15 25 1997

Murray & Nadel's Textbook of Respiratory Medicine E-Book Robert J. Mason, Arthur Slutsky, John F. Murray, Jay A. Nadel, Michael B. Gotway, 2015-03-17 Ideal for fellows and practicing pulmonologists who need an authoritative comprehensive reference on all aspects of pulmonary medicine Murray and Nadel s Textbook of Respiratory Medicine offers the most definitive content on basic science diagnosis evaluation and treatment of the full spectrum of respiratory diseases Full color design enhances teaching points and highlights challenging concepts Understand clinical applications and the scientific principles of respiratory medicine Detailed explanations of each disease entity allow you to work through differential diagnoses Expert Consult eBook version included with purchase This enhanced eBook experience offers content updates videos review questions and Thoracic Imaging Cases TICs all of which are easily navigable on any device for access on rounds or in the clinic Includes more than 1 000 figures and over 200 videos and audio files Key Points and Key Reading sections highlight the most useful references and resources for each chapter An expanded sleep section now covers four

chapters and includes control of breathing consequences of sleep disruption as well as obstructive and central apnea New chapters in the Critical Care section cover Noninvasive Ventilation NIV and Extracorporeal Support of Gas Exchange ECMO New chapters focusing on diagnostic techniques now include Invasive Diagnostic Imaging and Image Guided Interventions and Positron Emission Tomography and a new chapter on Therapeutic Bronchoscopy highlights the interventional role of pulmonologists Embedded videos feature thoracoscopy therapeutic bronchoscopy volumetric chest CT scans and more Brand new audio files highlight normal and abnormal breath sounds and the separate components of cough

ASE's *Comprehensive Echocardiography E-Book* American Society of Echocardiography, 2021-02-18 Edited by a team of leading echocardiography experts and endorsed by the American Society of Echocardiography ASE S Comprehensive Echocardiography 3rd Edition covers the full spectrum of sonography of the heart in one succinct authoritative resource This highly regarded text provides must know information on everything from basic foundations and principles to clinical application written and edited by ASE members with expertise in each specific area Case studies numerous tables high quality images and videos highlight the latest uses of echocardiography including the most recent 2D and 3D advances Discusses all the latest methods to assess cardiac chamber size and function valvular stenosis regurgitation cardiomyopathies coronary artery disease complications of myocardial infarction and other cardiac pathologies Covers recent advances in critical care echocardiography cardio oncology structural heart disease interventional intraoperative echocardiography strain imaging of left and right heart chambers multimodality imaging in systemic diseases and novel 3D techniques Contains more than 1 200 updated images echocardiograms including 2D 3D and Doppler diagrams anatomic drawings algorithmic drawings and more Provides access to nearly 600 full motion echocardiography video clips Keeps you up to date with the latest echocardiography practice guidelines and advanced technologies Enhanced eBook version included with purchase Your enhanced eBook allows you to access all of the text figures and references from the book on a variety of devices

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Reviewing **A Journey With Pulmonary Arterial Hypertension**: Unlocking the Spellbinding Force of Linguistics

In a fast-paced world fueled by information and interconnectivity, the spellbinding force of linguistics has acquired newfound prominence. Its capacity to evoke emotions, stimulate contemplation, and stimulate metamorphosis is truly astonishing. Within the pages of "**A Journey With Pulmonary Arterial Hypertension**," an enthralling opus penned by a highly acclaimed wordsmith, readers set about an immersive expedition to unravel the intricate significance of language and its indelible imprint on our lives. Throughout this assessment, we shall delve to the book is central motifs, appraise its distinctive narrative style, and gauge its overarching influence on the minds of its readers.

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A Journey With Pulmonary Arterial Hypertension Introduction

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