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in this issue of cardiology clinics guest editor richard a krasuski brings his considerable expertise to the topic of pulmonary hypertension top experts in the field cover key topics such as the pathobiology of pulmonary arterial hypertension pulmonary hypertension in hiv infection lung disease related pulmonary hypertension pulmonary hypertension in connective tissue disorders and more contains 12 relevant practice oriented

topics including surgical management of chronic thromboembolic pulmonary hypertension advanced circulatory support and lung transplantation in pulmonary hypertension interventional management of chronic thromboembolic pulmonary hypertension and more provides in depth clinical reviews on pulmonary hypertension offering actionable insights for clinical practice presents the latest information on this timely focused topic under the leadership of experienced editors in the field authors synthesize and distill the latest research and practice guidelines to create clinically significant topic based reviews

pulmonary hypertension is a fatal lung and heart disease it is characterized by shortness of breath fatigue and fainting it is exacerbated by an increase of the pressure in the lung vasculature through exercise leading to progressive worsening of hemodynamics right ventricular hypertrophy right heart insufficiency and finally right heart failure this book focuses on pulmonary arterial hypertension a rare and progressive subgroup of pulmonary hypertension which is today incurable and terminally faal classification of pulmonary arterial hypertension its pathology and strategies for future therapy will be of interest both to those suffering from the disease and those who take care of patients this volume will also engage physicians and other scientists contributing to an understanding of the pathophysiology of pulmonary arterial hypertension and attempting to extend life of humans with pulmonary arterial hypertension by developing causal and curative therapies

this issue of clinics in chest medicine guest edited by dr aaron b waxman and dr inderjit singh is focused on pulmonary hypertension topics discussed in this issue include but are not limited to integrative omics to characterize and classify pulmonary vascular disease contemporary pharmacotherapeutic approach in pulmonary arterial hypertension personalized medicine the future management of pulmonary hypertension requires a new taxonomy sex differences in pulmonary hypertension and pulmonary hypertension in pregnancy

pulmonary hypertension ph is a diverse group of diseases that elevates pulmonary artery pressure globally ph prevalence is approximately 1 pulmonary arterial hypertension incidence is 6 per million patients with a prevalence of 49 55 per million this book dives into historical facts related to ph clinical features treatment and specialized issues associated with ph this book is a resource for health professionals such as nurses medical students allied health professionals primary care physicians pulmonary clinicians and clinicians caring for patients with pulmonary hypertension

this book focuses on pulmonary arterial hypertension pah group 1 and chronic thromboembolic pulmonary hypertension cteph group 4 among the various groups of pulmonary hypertension ph whose classification was updated into five major categories at the 5th world symposium held in nice france in 2013 readers will find recent progress methods and up to date information on ph mechanisms diagnostic images and treatment in the management of ph this volume with contributions by leading researchers worldwide in the field consists of five parts starting with the fundamentals of ph then pathophysiology and genetics treatment and right ventricular function

pulmonary hypertension is a world wide problem which is gaining greater attention and is more frequently diagnosed now more than the past ten or twenty years because of the use and interpretation of echocardiograms pulmonary hypertension raises questions and discusses concepts which will address the issue of etiology of pulmonary hypertension its pathobiology with its close proximity to cancer and the consequences of severe ph which is right heart failure and death

guest editor terence k trow has assembled an expert team of authors on the topic of pulmonary arterial hypertension articles include epidemiology of pulmonary arterial hypertension pathology of pulmonary hypertension genetics of pulmonary arterial hypertension diagnosis of pulmonary arterial hypertension pulmonary hypertension owing to left heart disease pulmonary hypertension due to lung disease and or hypoxia pulmonary arterial hypertension associated with congenital heart disease world health organization group 5 pulmonary hypertension and more

pulmonary arterial hypertension pah is a relatively rare but potentially life threatening disease in most forms the disease is likely to be diagnosed late and is associated with progressive clinical deterioration and premature death a clinician s guide to pulmonary arterial hypertension second edition enhances the overall pah awareness of the

this monograph aims to provide an in depth overview of our current understanding of the various forms of pulmonary hypertension their diagnosis and their treatment

pulmonary arterial hypertension pah is a high blood pressure in the arteries that supply the lungs with blood patients with mild pah particularly young patients may feel quite well and get breathless or tired only with vigorous exertion those with severe pah may get breathless on minimal exertion and feel tired exhausted and experience chest pain forceful heart beats dizzy turns loss of appetite and have swelling of the legs due to fluid retention this book will help patients and their families understand what pah is and advise them on how to cope with their illness in the uk patients with pah are referred to one of the eight national pulmonary hypertension centres staffed by a team of specialist doctors nurses pharmacists and other staff who are experienced and expert in managing all aspects of the condition currently the drugs used to treat the condition can be prescribed only by these specialists the book will explain what the illness is and how it is treated in a reassuring and understandable way the book will also be of interest to gps nurses and other related healthcare professionals

pulmonary hypertension is defined as a mean pulmonary artery pressure mpap 25 mmhg with pulmonary capillary wedge pressure 15 mmhg measured by cardiac catheterization the etiology of ph has a varied spectrum extending right from drugs toxins and portal hypertension to hiv collagen vascular diseases and persistent pulmonary hypertension of newborn etc the estimation of disease prevalence has been nearly

impossible owing to the geographic distribution and economic diversity along with significant regional variations in human development and healthcare infrastructure a large number of patients with ph never reach the health centers capable of diagnosing the disease condition correctly advance pulmonary vascular disease as a result of uncorrected chd is a major health challenge in the developing world ph exists as a major component of many forms of cardiac and pulmonary disease while breathlessness is the most common feature of ph patients often also present with chest pain syncope fatigue weakness and abdominal distension the precordial signs include a right ventricular lift accentuated pulmonary component of s2 a pansystolic murmur of tricuspid regurgitation a diastolic murmur of pulmonary regurgitation and a right ventricular s3 the standard diagnostic workup in developed countries includes a series of investigations to rule out the secondary causes additional tests are required to estimate the disease severity and plan the appropriate treatment these include the cardiac catheterization selective pulmonary angiography by direct injection of pulmonary arteries high resolution ct scan cardiac magnetic resonance abgs nocturnal o2 saturation etc while most of the basic management is feasible in the indian conditions most of the newly introduced drugs are either not available or are available at costs that far exceed the paying capacity of an average citizen of a developing economy an underdeveloped health insurance system adds further to the financial burden of the treatment measures like formulation of guidelines for diagnosis and treatment of pah educating clinicians and scientists and making medications affordable to poor patients might ensue a breakthrough in the overall management of pulmonary hypertension

this book provides the framework for a singular reference in the field of pulmonary hypertension pulmonary vascular disease is a complex and heterogeneous condition characterized by remodeling of distal pulmonary arterioles that increases pulmonary vascular resistance to affect cardiopulmonary hemodynamic and right ventricular function adversely resulting in a clinical syndrome of diminished exercise tolerance shortness of breath and heart failure associated morbidity and mortality owing to the availability of novel pulmonary circulation selective pharmacotherapies over the previous decade the number of pulmonary hypertension patients eligible for treatment has increased substantially despite this progress under awareness persists within the practicing pulmonary cardiovascular and general internal medicine communities this is due in part to the complex array of molecular mechanisms implicated in the pathobiology of ph as well as cutting edge discoveries from translational scientific works that provide a new framework by which to understand pulmonary vascular right ventricular coupling taken together a key educational opportunity is exposed to bridge this knowledge gap through the synthesis of a contemporary text that emphasizes basic science translational and clinical principles and treatment strategies for understanding pulmonary hypertension

this book is intended as a comprehensive practically oriented reference on pulmonary hypertension within the context of adult congenital heart disease achd after an introductory chapter on pathophysiology the various types of pulmonary hypertension that may be encountered in achd are

discussed highlighting the specifics observed within different patient categories the diagnostic approach is then addressed in detail and the last section of the book is devoted to management options from conservative approaches to interventional treatment and the concept of treat and repair management in specific patient subjects such as pregnant women fontan patients and down syndrome patients with eisenmenger syndrome is fully discussed and guidance is also provided on palliative care pulmonary arterial hypertension related to congenital heart disease pah chd despite significant similarities in lung pathohysiology differs significantly from other types of pah in terms of mechanism of onset natural history and management mistakes and pitfalls in the management of patients with pah chd are often related to a lack of knowledge or expertise in this condition pulmonary hypertension in adult congenital heart disease will be a valuable resource and learning tool for all who care for patients with achd both in tertiary practice and general cardiology

this book is a clinical guide to controversial and emerging topics in pulmonary hypertension there are multiple challenges and unanswered questions encountered by clinicians that evaluate diagnose and treat patients with suspected or confirmed pulmonary vascular disease this book provides a deep dive into the diagnosis and therapeutics of pulmonary hypertension supported by the literature and balanced with personal clinical experience expert authors have chosen these specific topics to address issues where uncertainty and or controversy exists as well as highlight areas that are just being incorporated into clinical practice these topics include exercise pulmonary hypertension sickle cell disease and pulmonary hypertension and sarcoid pulmonary hypertension among many others chapters address the diagnostic and treatment dilemmas posed by these various clinical entities through literature review sharing of expert opinion and review of recent guidelines and their applicability to the multiple different nuanced presentations of pulmonary hypertension this is an ideal guide for pulmonologists cardiologists and other specialty practitioners caring for patients with pulmonary hypertension

first book dedicated to this disease previously thought to be incurable but with the advent of new drugs now amenable to management and a much improved prognosis for patients from the pah association the leading experts in field incorporates the latest aacp management guidelines includes evidence based treatment algorithms based on the recently updated accp guidelines for medical treatment aimed at specialists in pulmonology and cardiovascular disease this volume provides the clinician with the most up to date information on the effective management of pah

we are grateful for the support of actelion a sponsor of this research topic whose cooperation has contributed to fostering scientific discovery by reducing article publishing costs for some authors we hereby state publicly that actelion has had no editorial input in articles included in this research topic thus ensuring that all aspects of this research topic were evaluated objectively unbiased by any specific policy or opinion of actelion actelion is part of the johnson johnson family of companies we are leaders in the science and medicine of pulmonary arterial hypertension pah

with over 15 years of experience in this devastating cardiovascular disorder

this volume presents overviews as well as in depth reviews of many aspects of the clinical presentation pathophysiology and treatment of pulmonary hypertension ph especially ph related to thromboembolic disease saleem sharieff presents a comprehensive synopsis of the epidemiologic clinical histopathologic and therapy of pah next dimitar sajkov bliegh mupunga jeffrey j bowden and nikolai petrovsky comprehensively review world health organization group iii ph the cellular and biochemical pathophysiology of ph are summarized by rajamma mathew specific mechanisms implicated in the pathogenesis of ph are presented by junko maruyama ayumu yokochi erquan zhang hirohumi sawada kazuo maruyama and aureliano hernandez and rafael a areiza jean elwing and ralph panos discuss ph associated with acute thromboembolism mehdi badidi and m barek naz discuss ph caused by chronic thromboembolic disease juan c grignola maria j ruiz cano juan p salisbury gabriela pascal pablo curbelo and pilar escribano present the physiologic assessment of patients with chronic thromboembolic disease prior to surgical pulmonary endarterectomy and finally henry liu philip l kalarickal yiru tong daisuke inui michael j yarborough kavitha a mathew amanda gelineau and charles fox comprehensively review the clinical perioperative evaluation and management of patients with ph due to chronic thromboembolic disease

this concise pocketbook provides an easily accessible resource on pulmonary arterial hypertension pah for medical professionals senior and trainees nurses and allied disciplines pah is not any longer an orphan disease nor is it associated with a grave prognosis and premature death as it used to be the case a decade or two ago patients with pah should enjoy improved survival and quality of life provided that an early and not late diagnosis is made combined with timely initiation of advanced therapy in specialized designated tertiary centres this comprehensive text incorporates pah expertise from the uk and the rest of the world the book outlines the key points with respect to the latest classification pathobiology genetics clinical assessment of the patient with suspected pah and the role of imaging there are specific chapters addressing different pah aetiologies namely idiopathic pah thromboembolic pah pah related to connective tissue disease congenital heart disease eisenmenger complex respiratory disease and other unusual causes last but not least the book addresses counselling contraception and the latest therapy for the challenging area of pregnancy and pah which is still associated with a high maternal mortality risk the main objective of the book is to increase awareness of pah promote rapid diagnostic work up and timely specialist referral so that effective therapy is made available as early as possible to all patients with suspected or known pah physicians senior or junior nurse or other health care professional whether senior or junior who may encounter patients with pah has much to gain from this book

pulmonary hypertension ph is increased pressure in the pulmonary arteries which carry blood from the heart to the lungs to pick up oxygen the changes resulting from ph make it difficult for the heart to push blood through the pulmonary arteries causing the heart to become weak and

possibly to develop failure understanding the causes and treatment of ph can help heart failure specialists prevent heart failure due to ph

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