

Case Report

Growing up with Idiopathic Pulmonary Arterial Hypertension: An Arduous Journey

Tanguy Dutilleul ¹, Nesrine Farhat ² , Ruth Heying ³, Marie-Christine Seghaye ^{2,*}  and Maurice Beghetti ⁴ 

¹ Department of Pediatric Cardiology, University Hospital Aachen, 52074 Aachen, Germany

² Department of Pediatric Cardiology, Centre Hospitalier Universitaire de Liège, 4000 Liège, Belgium

³ Department of Pediatric Cardiology, University Hospital Leuven, 3000 Leuven, Belgium

⁴ Department of Pediatric Cardiology, Children's University Hospital Geneva, 1205 Geneva, Switzerland

* Correspondence: msegghaye@chuliege.be; Tel.: +32-4-323-9275

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

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A Journey With Pulmonary Arterial Hypertension:

This book delves into A Journey With Pulmonary Arterial Hypertension. A Journey With Pulmonary Arterial Hypertension is an essential topic that needs to be grasped by everyone, from students and scholars to the general public. The book will furnish comprehensive and in-depth insights into A Journey With Pulmonary Arterial Hypertension, encompassing both the fundamentals and more intricate discussions.

1. This book is structured into several chapters, namely:
 - Chapter 1: Introduction to A Journey With Pulmonary Arterial Hypertension
 - Chapter 2: Essential Elements of A Journey With Pulmonary Arterial Hypertension
 - Chapter 3: A Journey With Pulmonary Arterial Hypertension in Everyday Life
 - Chapter 4: A Journey With Pulmonary Arterial Hypertension in Specific Contexts
 - Chapter 5: Conclusion
 2. In chapter 1, the author will provide an overview of A Journey With Pulmonary Arterial Hypertension. The first chapter will explore what A Journey With Pulmonary Arterial Hypertension is, why A Journey With Pulmonary Arterial Hypertension is vital, and how to effectively learn about A Journey With Pulmonary Arterial Hypertension.
 3. In chapter 2, this book will delve into the foundational concepts of A Journey With Pulmonary Arterial Hypertension. This chapter will elucidate the essential principles that must be understood to grasp A Journey With Pulmonary Arterial Hypertension in its entirety.
 4. In chapter 3, this book will examine the practical applications of A Journey With Pulmonary Arterial Hypertension in daily life. This chapter will showcase real-world examples of how A Journey With Pulmonary Arterial Hypertension can be effectively utilized in everyday scenarios.
 5. In chapter 4, this book will scrutinize the relevance of A Journey With Pulmonary Arterial Hypertension in specific contexts. The fourth chapter will explore how A Journey With Pulmonary Arterial Hypertension is applied in specialized fields, such as education, business, and technology.
 6. In chapter 5, the author will draw a conclusion about A Journey With Pulmonary Arterial Hypertension. This chapter will summarize the key points that have been discussed throughout the book.
- This book is crafted in an easy-to-understand language and is complemented by engaging illustrations. This book is highly recommended for anyone seeking to gain a comprehensive understanding of A Journey With Pulmonary Arterial Hypertension.

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A Journey With Pulmonary Arterial Hypertension Introduction

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