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Overview of the Most Prevalent Pediatric Congenital Heart Diseases: A Literature Review

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Author's contribution

The sole author designed, analysed, interpreted and prepared the manuscript.

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Review Article

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ABSTRACT

Like every other prevalent disease process, cardiovascular diseases pose a great risk for the morbidity and mortality of human beings. Pediatric cardiovascular disease (CVD) represents a broad spectrum of disorders that affect both the heart and blood vessels in the pediatric population. There have been several epidemiological studies done to highlight the prevalence and incidence of various pediatric CVDs, emphasizing the impact on global child health. Notably, congenital heart defects, acquired heart diseases, and cardiomyopathies contribute to the majority of pediatric CVDs. Understanding the prevalence and distribution of these conditions is important for resource allocation and healthcare planning.

Similarly, several risk factors contribute to the development and progression of pediatric CVD. Genetic predisposition, prenatal exposures, environmental factors, and lifestyle choices all play pivotal roles. Identifying and addressing these risk factors is essential for early intervention and preventive strategies.

However, advances in diagnostic modalities have revolutionized the assessment of pediatric CVD. Non-invasive imaging techniques, genetic testing, and biomarker analyses enable earlier and more accurate detection, facilitating timely intervention and personalized treatment plans. Additionally,

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emerging therapeutic approaches, including innovative medical interventions and surgical techniques, are transforming the landscape of pediatric CVD management. This abstract aims to serve as a concise resource for clinicians, researchers, and physicians interested in pediatric cardiovascular health. By understanding the epidemiology, recognizing key risk factors, and staying aware of the emerging trends in diagnosis and management, people can contribute to improved outcomes and enhanced quality of life for children affected by cardiovascular diseases.

Keywords: Cardiovascular diseases; pediatric cardiology; pediatric CVD; cardiovascular mortality; pediatrics.

1. INTRODUCTION

Pediatric cardiovascular diseases (CVD) constitute a complex and diverse range of conditions that affect the hearts and blood vessels of children. The prevalence of pediatric CVD, that includes congenital anomalies, acquired disorders, and cardiomyopathies, highlights the significance of understanding this intricate medical landscape. As global health priorities continue to evolve, there is a growing recognition of the need for comprehensive insights into the epidemiology, risk factors, and emerging trends associated with pediatric CVD [1].

In recent years, considerable progress has been made in unraveling the complexities of pediatric CVD. Epidemiological studies have shed light on the prevalence and distribution of these conditions, emphasizing the global burden on pediatric health. From congenital heart defects to acquired cardiac conditions, the spectrum of pediatric CVD demands a comprehensive understanding to inform healthcare planning and resource allocation [2].

The etiology of pediatric CVD is multifactorial, involving a delicate interplay of genetic predispositions, prenatal influences, environmental factors, and lifestyle choices. Recognizing and addressing these diverse risk factors are pivotal for developing effective prevention and intervention strategies. Moreover, advancements in diagnostic tools and therapeutic modalities have transformed the landscape of pediatric CVD care, allowing for earlier detection and more personalized treatment approaches [3].

In the search to develop a better understanding of different pediatric cardiovascular diseases, we can see that currently, there is a specialized branch that revolves around anatomical malformations and has significantly marked its evolution throughout [4].

Firstly, subsequent to substantial advancements in pediatric cardiac surgery during the latter half of the previous century, a myriad of interventional techniques has emerged. These techniques efficiently address various structural issues without necessitating open cardiac surgery and extra cardiac circulation [5].

Secondly, in recognizing that perioperative survival now serves as a quality criterion rather than merely an “achievement,” the current challenge lies in extending our focus to include middle- and long-term survival and morbidity. The goal is to guide individuals within the “congenital heart disease population” toward reaching their eighties in optimal health. This transformative shift redirects research emphasis from operative techniques to perioperative management and long-term continuous care. Achieving this necessitates a profound understanding of physiological, systemic pediatric and pharmacological nuances [6].

Thirdly, novel echocardiographic techniques and advancements in cardiac MRI have allowed for the introduction of a new era of improved assessment of systolic and diastolic function, synchrony, tissue characterization, and flow measurements [7].

Overall, this progress enables detailed anatomical and functional evaluations, potentially paving the way for a more nuanced and personalized therapeutic approach. In short, the landscape of pediatric cardiology has evolved to not only address structural anomalies through innovative interventions but also to extend its purview towards ensuring sustained well-being and personalized care for the congenital heart disease population [8].

2. AN OVERVIEW OF THE MOST PREVALENT PEDIATRIC CARDIO-VASCULAR DISEASES

In no particular order of preference, given in the preceding section is an overview of the most

prevalent pediatric cardiovascular diseases in society today. These pediatric CVDs have been affecting millions of infants all around the world. Despite adequate monitoring, evaluation, and prior diagnostic approaches present, little could be done to contain the escalating situation.

2.1 Tetralogy of Fallot (TOF)

Tetralogy of Fallot (ToF) stands as one of the most prevalent cyanotic congenital heart anomalies, characterized by a quartet of distinctive features: a ventricular septal defect (VSD), a typically dynamic right ventricular (RV) outflow tract obstruction (RVOTO), an overriding aorta, and right ventricular hypertrophy (RVH) [9].

This condition accounts for a substantial 7 - 10% of all congenital cardiac malformations, occurring at an incidence of approximately one in 3,500 live births [10].

The recognition of ToF as a series of malformations dates back to 1671; however, it acquired its name from the French physician Etienne-Louis Fallot [9].

In 1888, Fallot presented the first case series, providing detailed anatomical and pathological descriptions of the four cardinal features.

2.2 Ventricular Septal Defect (VSD)

Typically, this condition manifests as a sizable, non-restrictive defect, indicating an absence of obstruction to the flow across the ventricular septal defect (VSD) [11].

The VSD is commonly situated in the perimembranous and muscular regions of the ventricular septum, facilitating the shunting of blood between the ventricles. In the classical cyanotic Tetralogy of Fallot (ToF), this results in a right-to-left blood flow pattern [12].

The anatomical positioning of the VSD plays a crucial role in the pathophysiology, enabling the characteristic hemodynamic changes associated with this congenital heart anomaly [13].

2.3 Dynamic Right Ventricular Outflow Tract Obstruction (RVOTO)

This obstruction, termed RVOTO, plays a crucial role in the development of another hallmark feature of ToF - right ventricular hypertrophy (RVH). In the presence of an unrestrictive ventricular septal defect (VSD), escalating RVOTO amplifies right ventricular (RV) pressure. Consequently, a right-to-left shunt is driven through the VSD, resulting in diminished pulmonary blood flow and subsequent hypoxemia [14].

In cases where the patent ductus arteriosus (PDA) is closed and no collateral supply exists, the severity of RVOTO becomes a determining factor in the degree of hypoxemia observed in the patient upon presentation [15].

The more pronounced the RVOTO, the more profound the hypoxemia, emphasizing the critical interplay between anatomical features and hemodynamic consequences in the complex pathophysiology of Tetralogy of Fallot [16].

2.4 Overriding Aorta

This condition delineates the ventriculoarterial connection, where the aorta exhibits variable overriding of the ventricular septal defect (VSD) due to malalignment of the outlet component of the septum. In Tetralogy of Fallot (ToF), the aorta predominantly originates from the left ventricle, with only a partial origin from the right ventricle - an occurrence referred to as the “override.” This overriding nature is a distinctive feature [9].

Should the aorta predominantly arise from the right ventricle, the anomaly is termed double-outlet right ventricle (DORV). The physiological implications are contingent on the position of the VSD and any associated right ventricular (RV) or left ventricular (LV) outflow obstruction. In essence, the nature of the ventriculoarterial connection, characterized by the degree of aortic override and the associated anatomical variations, significantly influences the overall hemodynamics and clinical manifestations in these congenital heart conditions [17].

2.5 Right Ventricular Hypertrophy (RVH)

The development of right ventricular hypertrophy (RVH) arises as a direct consequence of right ventricular outflow tract obstruction (RVOTO). The increased pressure within the right ventricle (RV) is a compensatory mechanism essential for sustaining pulmonary blood flow in the presence of an obstruction. Consequently, the persistent elevation in RV pressure leads to structural changes; affecting both the size and muscle mass of the RV [18].

These alterations in RV cavity size and muscle mass hold critical significance, particularly in the post-repair phase following Tetralogy of Fallot (ToF) correction. The surgical intervention aims to address the anatomical anomalies associated with ToF, including the RVOTO [19].

Through the lens of historical and clinical understanding, the Tetralogy of Fallot remains a noteworthy condition, both in its prevalence among congenital cardiac malformations and the enduring legacy of its eponymous identifier, Etienne-Louis Fallot.

In its early stages, when confronted with profound cyanosis, interventions aimed at alleviating the condition involved establishing pulmonary blood flow through systemic-to-pulmonary shunts. This marked the initial steps in the evolution of surgical strategies.

Over time, there has been a transformative progression in surgical repair techniques, including innovative approaches such as pulmonary valve-sparing techniques. The field has witnessed a trend toward corrective operations being performed at increasingly younger ages, reflecting advancements in both surgical expertise and perioperative care.

2.6 Patent Ductus Arteriosus (PDA)

The ductus arteriosus serves as a vital fetal vessel facilitating the bypass of oxygenated blood from the placenta, allowing it to circumvent the fetal lungs during gestation [20].

Upon birth, a newborn takes its first breath, initiating the inflation of the lungs with air. This respiratory shift leads to a drop in pulmonary vascular resistance, enabling blood to flow from the right ventricle to the lungs for oxygenation [21].

The increase in arterial oxygen tension, coupled with reduced blood flow through the ductus arteriosus, triggers its constriction and functional closure within 12 to 24 hours of age in healthy, full-term newborns. Anatomically, permanent closure typically occurs within 2 to 3 weeks after birth [22].

However, in premature infants, the closure of the ductus arteriosus does not progress as rapidly. This delay may necessitate pharmacologic or surgical interventions to achieve closure and address potential unwanted repercussions.

The management of the ductus arteriosus is crucial in optimizing the transition from fetal to neonatal circulation, particularly in premature infants who may require additional medical attention to support their cardiovascular adaptation post-birth [23].

2.7 Arrhythmias

Pediatric arrhythmias hold significant importance within the field of pediatric cardiology. Undoubtedly, this field has also attracted significant attention from pediatric electrophysiologists due to advancements in diagnostic and treatment methods in recent years [24].

The classification of pediatric arrhythmias generally falls under three primary headings: tachyarrhythmias, bradyarrhythmias (including sinus node dysfunction and atrioventricular (AV) blocks), and primary ion channel defects, commonly referred to as channelopathies [25].

AVRT, or Atrioventricular Reentrant Tachycardia, is alternatively referred to as accessory-pathway mediated tachycardia or orthodromic reciprocating tachycardia. It stands out as the predominant type of supraventricular tachycardia (SVT) observed in children, constituting a significant 82% of arrhythmias noted during infancy. This form of tachycardia involves an abnormal accessory pathway that allows electrical signals to circulate between the atria and ventricles, contributing to episodes of rapid heartbeat [26].

The majority of patients with pediatric arrhythmias can be accurately diagnosed, effectively treated, and monitored by pediatricians. Consequently, it is essential for every pediatrician encountering patients with pediatric arrhythmias to possess comprehensive knowledge about the associated diseases, discern ECG findings, and be well-versed in the available therapeutic options [27].

2.8 Infective Endocarditis

Infective endocarditis (IE) is an uncommon condition, particularly rare in children, with an estimated incidence ranging from 0.43 to 0.69 cases per 100,000 children in a year [28].

This low frequency poses challenges in gathering substantial evidence to establish optimal

diagnostic and therapeutic approaches for IE in pediatric populations.

Globally, the occurrence of IE in children is lower than in adults. However, a notable increase has been observed in recent decades, attributed to the heightened prevalence of risk factors such as congenital heart disease or the use of intravascular devices, such as central catheters, pacing leads, etc [29].

Children diagnosed with IE constitute a group characterized by a notable rate of events during follow-up. These events enforce the need for reinterventions and recurrent episodes of IE. Although congenital heart disease remains the most prevalent risk factor, there is a rising frequency of other factors associated with nosocomial IE.

3. CONCLUSION

In conclusion, pediatric cardiovascular diseases (CVD) encompass a diverse spectrum of conditions affecting the hearts and blood vessels of children. While congenital heart defects remain a significant contributor to the pediatric CVD landscape, acquired heart diseases and cardiomyopathies add complexity to the diagnostic and therapeutic challenges faced by healthcare professionals. In the field of pediatric CVD, collaboration and continued research are pivotal in addressing the challenges posed by these conditions. By staying aware of emerging trends, and innovative technologies, and fostering interdisciplinary cooperation, the medical community can strive towards furthering the well-being of children affected by cardiovascular diseases.

CONSENT

It is not applicable.

ETHICAL APPROVAL

It is not applicable.

COMPETING INTERESTS

Author has declared that no competing interests exist.

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