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It is the official journal of the Global Forum on Humanitarian Medicine in Cardiology and Cardiac Surgery (GFHM) whose Mission is to close the disparity between the medical care facilities of the North and those of the South by developing programs & projects related to education, research, facilities, quality improvement and training in these countries.

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Bicuspid Aortic valve: a frequent syndrome with a challenging outcome

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Editorial

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Abbreviations

BAov: Bicuspid Aortic valve; CHD: Congenital Heart Disease; HLHS: Hypoplastic Left Heart Syndrome; PDA: Patent Ductus Arteriosus; VSD: ventricular septal defect; CoA: Coarctation of the Aorta; CVS: cardiovascular system; AAo: Ascending Aorta; TTE: Transthoracic Echocardiography; TOE: Trans Oesophageal Echocardiography; AVR: Aortic Valve Replacement; TAVI: Transcatheter Aortic Valve Implantation.

The Bicuspid Aortic valve (BAov) has the ability not only being among the most common congenital heart diseases (CHD) but also having both a fascinating clinical presentation and a challenging outcome. It can be found in approximately 1-2% of the general population and mostly among males. When found in females it is mostly seen in severe clinical cases. This condition can present in any age from fetal to late adulthood, even...never! It has a variety of clinical spectrum that can range from the extreme form of a Hypoplastic Left Heart Syndrome (HLHS) to undetected minimal valvular disease- stenosis or/and regurgitation - or incidentally present as a sub-clinical endocarditis; only mentioning a few of its many presentations. As its adverse cardiovascular outcomes are more common than previously thought, therefore due to its high prevalence it represents a huge burden towards public cardiovascular care.

Despite Leonardo's da Vinci first description of the defect, and Sir William Osler's approach as the most common CHD, we still have more questions regarding the disease than answers¹.

Up to date, genetic causes, and clinical implications for the majority of BAov patients remain largely unknown. Possibly a genetic component exists, leading to a link to other CHD's such as Patent Ductus Arteriosus (PDA), ventricular septal defect (VSD) and coarctation of the aorta (CoA). Evidence of an autosomal dominant inheritance pattern with variable expression and incomplete penetrance in families has been described. Additionally, mutations in NOTCH1 - a mechanosensory receptor found in arteries - may associate with a BAov and valvular calcium-deposition reactivation. A familiar model involving specific mutations or distinction in GATA-5 - a transcription factor that regulates the proper embryological development of the cardiovascular system (CVS) - have been linked to specific cases of BAov and aortopathy. A multifactorial event during the embryogenesis of the semilunar valves, leading to a formation of a fusion between the aortic cusps, creating the defect, has been proposed by few researchers in the field².

Schaefer, et al. in 2008 described four subtypes of BAov: I. Fusion seen between right - left

cusps (80% cases), II. Fusion seen between right - noncoronary cusps (19% cases), III. Approximately 1% of cases with fusion between left - noncoronary cusps. (Figure 1).

An additional IV. Sub-type known as a “functional BAov”, where 3 uneven cusps can be seen with the valve closed but only two functioning, when opening, creating an echocardiographic image of a so called “fish mouth”³.

These can be identified in every day clinical assessment by 2D transthoracic echocardiography and in more detailed imaging in 3D. (Figure 2).

True BAov have only two symmetrical leaflets and two sinuses of Valsalva (Figure 2). Clinically, sub-type I is more likely to suffer stenosis in adults while subtype II valves will present pathology in younger ages (Figure 1)⁵.

Regarding the clinical presentation of a BAov we need to distinguish: A. a form pattern and B. an age pattern⁶. And although a “**silent**” form of BAov can exist and only be found as an incidental post-mortem finding the most common clinical presentation of a BAov is that of a as a **robust valvular disease**⁷.

A. Defying a form pattern; this indicates two common forms. Firstly, a **mostly stenosis** and a second **mixed form** where **regurgitation of the valve annulus** and **secondary aneurysm formations** of the AAO and augmented aortic dissection risk in nearby future can be expected.

The valve stenosis form may present from birth and/or gradually, as aging increases its amount of stenosis. In this form most probable in future the need for aortic valve replacement (AVR) is anticipated. Studies have shown this in 25-65% of patients in their late twenties age pe-

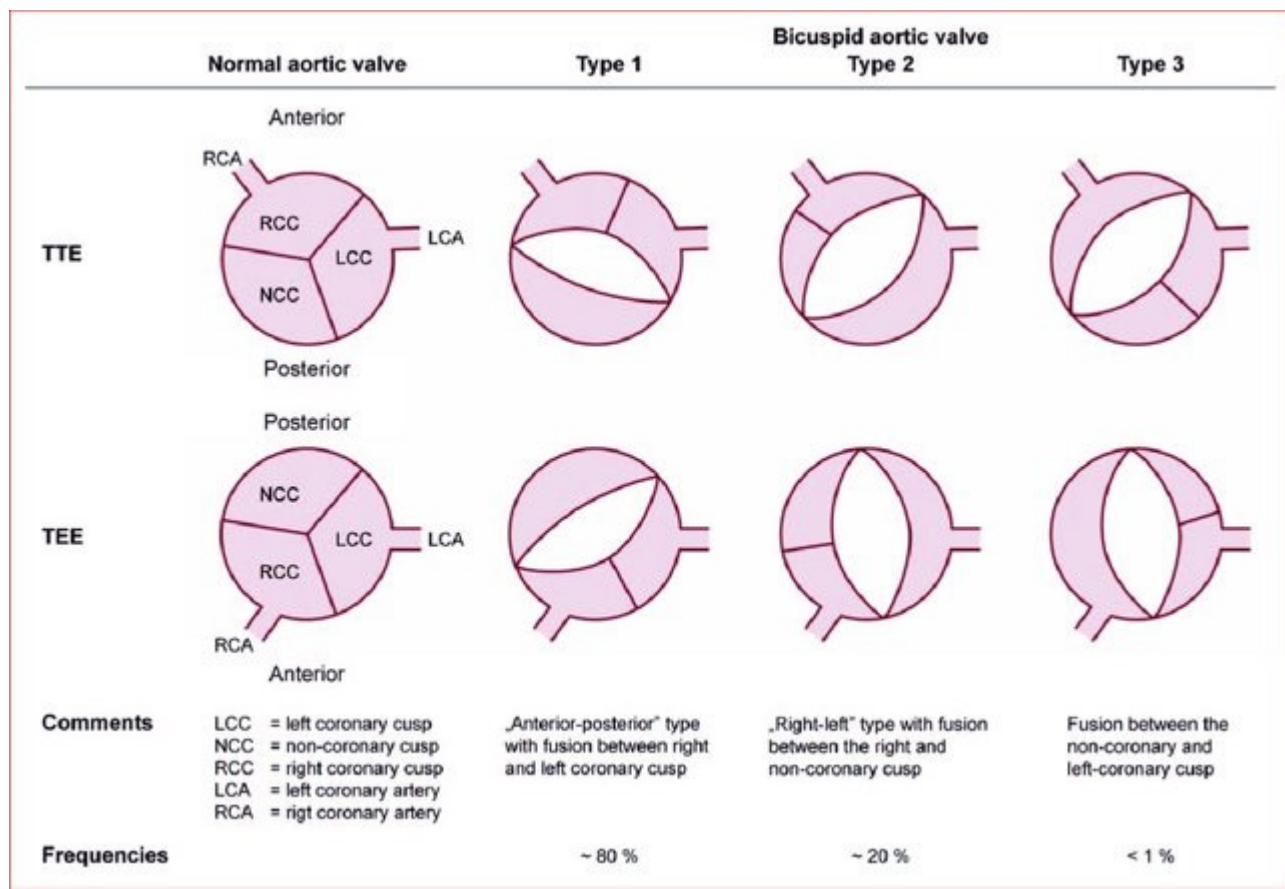


Figure 1: Anatomical classification of BAov. Courtesy of Prof. Christian R Habermann © From “Aortic aneurysms after correction of aortic coarctation: A systematic review”; February 2010; VASA.: Zeitschrift für Gefäßkrankheiten. Journal for vascular diseases 39(1):3-16

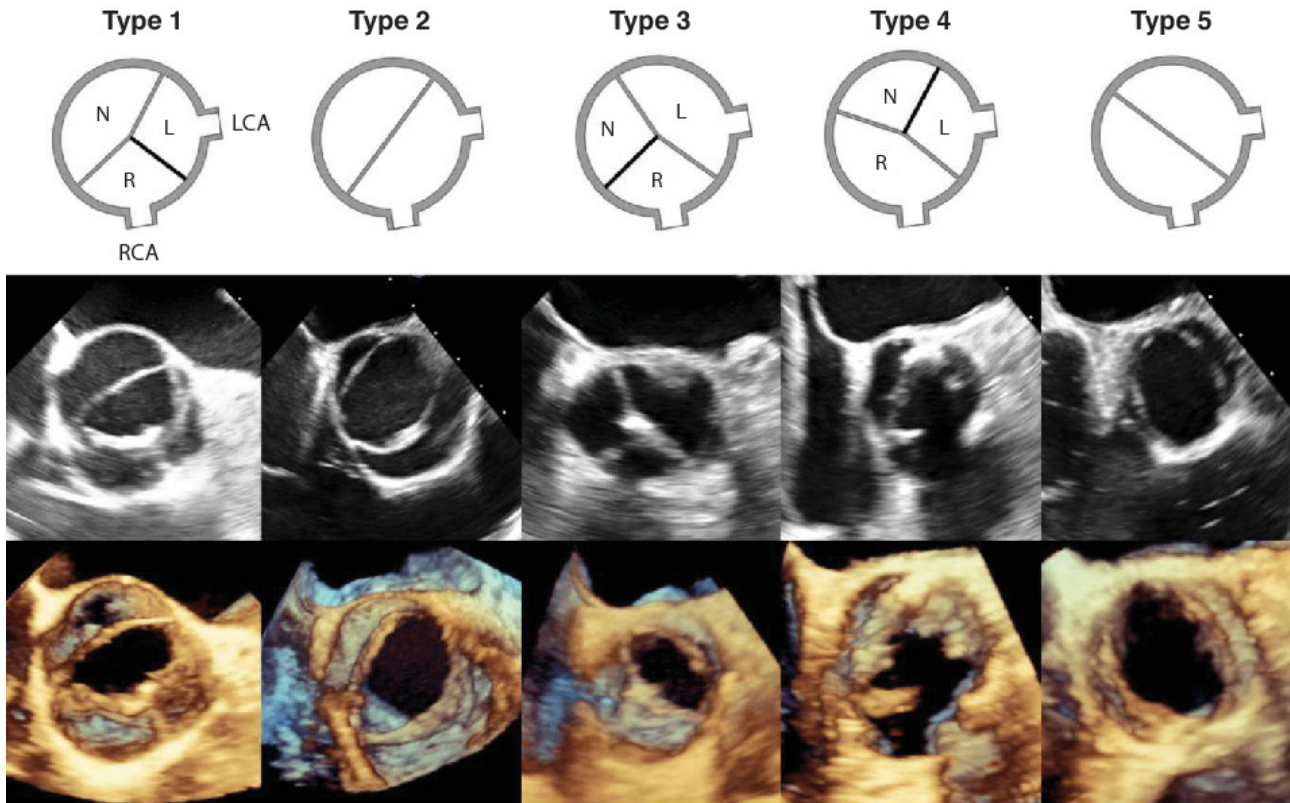


Figure 2: Courtesy of Prof. Selcen Yakar Tuluçe from his paper Assessment of bicuspid aortic valve phenotypes and associated pathologies: A transesophageal echocardiographic study. Turk Kardiyoloji Dernegi arsivi :1 December 2017

riod.

In the second form, studies have proven that 15-30% will need surgical repair of the BAovR+ between the 4th - 5th decade of their life. 45% of the group will suffer aneurysm formations while 26% will need to be operated in this age group.

Additional to the only or mostly valve clinical presentations a more **generalized aortopathy** form is also frequently seen among individuals suffering from a BAov.

Genetic evidence in combination with histological tissue alternations- because of cellular structural abnormalities including decreased fibrillin, causing smooth muscle cell detachment, and cell apoptosis. This element in collaboration to altered flow pattern abnormal jet orientation -due to uneven shear stress forces on the endothelium of the ascending aorta- because of two rather than three cusp, valve geometry, creates the setting of an aortopathy with multiple clinical

presentations. These clinical conditions involve most commonly the tubular AAO, the entire AAO, including the sinuses of Valsalva and sinotubular junction. Finally, in sub-type I BAov and male gender frequently exhibit dilatation of the area of the sinuses of Valsalva.

B. Defying the age pattern; a “**pediatric form**”, presenting earlier in life. A large study from autopsy specimens, calculated a prevalence of 6.7% of these complex CHD including a BAov as one of their elements. Examples are: BAov with: CoA (51.5%), with VSD (20.5%), with CoA and VSD, with PDA. BAov has been also reported as a part of transposition of the great arteries (1%), HLHS, complete atrioventricular canal defect, Ebstein’s anomaly, partial or total anomalous pulmonary venous drainage, tetralogy of Fallot, double-outlet right ventricle, left ventricular septal diverticulum. It can also be a part of genetic syndromes that involve CHD. Such are Turners (30%), William’s, Down’s, Marfan’s,

and Loey-Dietz^{1,5,6}. These patients also present with a higher incidence of left dominance coronary artery pattern (57%) with higher incidence of immediate bifurcation of the left main coronary artery, and higher incidence of mean length of the left main coronary significantly shorter up to 10mm. Anomalous origins of both right and left coronary arteries and the origin of the left circumflex artery as well as a single left coronary artery, have been reported. Spontaneous coronary artery dissection may occur also. An “**adult form**”; in which: isolated stenosis (36%), regurgitation (44%), both (20%), worsening by age at least and possible by accumulation of cardiovascular risk factors that accelerate atherosclerosis, are common clinical presentations. Additional to these more “silent” clinical presentations, formation of aneurysms (17-45%), rupture of them (10%), endocarditis (0.3-2%/yearly risk) and thrombotic events have been reported^{1,6}.

The natural history of BAov has been evaluated in several studies. It is known to be variable and dependent on associated abnormalities, age of patient and anatomical subtypes.

Diagnosis is based on clinical suspicion by medical, family history or incidentally finding during a well-baby or physical examination. Late presenters are common, mostly with “isolated-adult” valvul form. The mainstay of diagnosis is echocardiography (TTE or TOE) which can provide diagnosis in most patients also detecting progression of aortopathy. Recently, the use of metalloproteinase plasma assays, computer tomography and magnetic resonance imaging, have been introduced in clinical practice diagnosis and risk stratification^{1,6}.

Decisive treatment is only by surgical means. Medicines as: b-blockers, ACE-I, ARDS, and Statins, have been used to alleviate symptoms and slow progression. The 2017 ESC and the 2020 ACC/AHA guidelines for the management of patients with valvular heart disease address all the indications, types of surgery and/or intervention strategies in adult patients. The pediatric world still suffers from a lack of guideline; although clinicians in this field are encouraged to consult on the bases of the adult above mentioned guidelines^{6,8}.

In the childhood population valve replacement is not encouraged as physical development, outgrows the prosthetic valve. Strategies to repair the stenotic or regurgitant valve or even the use of a sub-coronary Ross procedure - by an experienced surgeon- replacing the affected BAov with their own pulmonary valve and using a pulmonary homograft and the side of the extracted valve, should be used because of the excellent postoperative long-term results^{9,10}. As valve calcification in children is absent a balloon valvuloplasty procedure is possible and can be in some cases the strategy of choice. Studies have shown good follow-up in both the immediate and medium terms, follow-up. This can buy time bridging the patient before needing surgery¹¹.

For the adult population a variety of surgical approaches can address any complex clinical presentation combining surgical repair, a Ross procedure, a biological and/or mechanical valve replacement with a Bentall procedure and/or more sophisticated vascular surgical procedures such as a Tairon David or a Magdi Yacoub procedure¹². Finally, for the elderly and complicated with additional extra cardiac nosology patients, a transcatheter aortic valve replacement approach (TAVI) can be offered⁹.

In summary, there are still large gaps in understanding the pathophysiology of **BAov-associated valvopathy - aortopathy**. Today, finding of a BAov in any age patient, must be approached as a **Syndrome** and not a defined disease.

As a **take home message**, the only robust data about the **syndrome of the BAov** is that we still have many to learn in how to deal efficiently with a condition in which its high prevalence, many subtypes and clinical presentations in any age group highlights it as a **major public health issue** in the field of congenital heart disease^{1,9}.

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The South American Paradigm of Congenital Heart Disease Care

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Abstract

South America is a land of contrasts. These contrasts encompass both nature and human activity. The health of individuals takes place within this context. In particular, congenital heart diseases, due to the complexity of their diagnosis and treatment, present a significant challenge for the region. Despite a large portion of the population lacking access to these services, the region is a pioneer in innovation in this field. Great surgeons from there have left an indelible legacy that still impacts the lives of people around the world today.

South America is a land of striking contrasts and breathtaking landscapes, featuring mountains, lakes, glaciers, waterfalls, islands, beaches, and rainforests teeming with wildlife. It is a vast and diverse continent with various cultures, languages, and ways of life. When studying the region, researchers often focus on the indigenous peoples who have lived there for thousands of years, as well as the societies that emerged during and after colonization.

One of the most remarkable aspects of South America is its cultural diversity. It is home to 440 million people and more than 400 indigenous groups, each with their languages, customs, and traditions. From the Quechua and Aymara peoples in the Andes to the Yanomami and Kayapo in the Amazon rainforest, these groups have strong connections to their land and close-knit communities. They have developed social structures that govern important aspects of life, including marriage, raising children, managing resources, and resolving conflicts.

The region's colonial history has also played a significant role in shaping its ethnographic landscape. Spanish and Portuguese explorers arrived in the 16th century, bringing new languages, religions, and ways of life. Over time, intermarriage between European settlers and indigenous peoples created a mix of cultural traditions and identities. Today, many South American countries have a blend of indigenous, European, and African influences, resulting in unique customs and traditions that reflect their complex histories.

In addition to the traditional cultures, South America is also home to bustling cities that are rapidly modernizing. Places like São Paulo, Buenos Aires and Lima are centers of innovation, technology, and entrepreneurship, attracting people from around the world. South America is a fascinating and dynamic region with a rich ethnographic landscape that continues to evolve over time.

The economic and financial situation of South America as a region is characterized by a mix of opportunities and challenges. While the region possesses abundant natural resources, including minerals, agricultural products, and energy

reserves, economic growth has been uneven across countries. Some nations have experienced significant progress, driven by sectors such as mining, agriculture, and manufacturing, while others face economic instability, high levels of inequality, and vulnerability to external shocks. Challenges such as political instability, corruption, inadequate infrastructure, and limited access to credit and investment hinder the region's overall economic development. Efforts to address these challenges and promote sustainable growth through regional cooperation, diversification of economies, and strengthening of institutions are crucial for fostering a more stable and prosperous economic future for South America.

Health systems and epidemiology of congenital heart defects in South America

Healthcare systems in South America are uneven and fragmented. There are four main sub-sectors: public health (governmental), private health, mandatory health, and direct out-of-pocket payments. The largest one is the public health system, which is primarily financed by the state. In this system, the federal or regional authority defines the budget allocated for healthcare services. It aims to provide universal and free access to healthcare for the entire population, ensuring that individuals are eligible to receive medical services at all levels of the healthcare system. However, due to the large population and limited resources, public health centers in countries like Argentina, Brazil, and Venezuela often face challenges with overburdened facilities and long waiting times for medical care.

Private health insurance operates on a voluntary basis, where individuals can purchase private insurance plans from commercial providers. Private health insurance offers access to a network of private healthcare providers and facilities. The quality and coverage of services depend on the specific insurance plan chosen and the individual's ability to afford the premiums.

Mandatory health insurance is another sub-system in South America. It typically requires individuals to contribute to a mandatory

health insurance fund, either through payroll deductions or other means. This system aims to ensure that all individuals have access to healthcare services, and the funds collected are used to finance the healthcare system. The coverage and benefits provided by mandatory health insurance can vary depending on the country and the specific regulations in place.

Lastly, there is direct out-of-pocket payment, which refers to individuals paying for healthcare services directly at the time of service without insurance coverage or mandatory contributions. This system is prevalent among individuals who do not choose the public sector, do not have access to private health insurance, or cannot contribute to a mandatory health insurance fund. Typically, access to this system is limited to individuals with higher incomes.

These four health systems -public health (governmental), private health insurance, mandatory health insurance, and direct out-of-pocket payment- represent different approaches to healthcare financing and access in South America, each with its advantages and challenges.

The diagnosis and treatment of congenital defects in South America occur within this context and exhibit disparities and fragmentation. This means that access to specialized care for congenital defects can vary widely across the region, leading to disparities in healthcare outcomes for affected individuals. Factors such as geographical location, socioeconomic status, and healthcare infrastructure contribute to these disparities. In some areas, individuals with congenital defects may face challenges accessing appropriate diagnostic tools, specialized medical professionals, and comprehensive treatment options.

In 2011, Kreutzer and colleagues conducted a comprehensive analysis of the epidemiology of congenital heart diseases (CHD) and pediatric cardiac surgery in South America¹. The study provided an overview of the annual birth rates in different countries within the region. Brazil had the highest birth rate, with approximately 3.7 million births, followed by Colombia with 800,000, and Argentina, Peru, Venezuela, Ecuador, Bolivia, and Chile each had around 700,000 births.

Paraguay, Uruguay, and Guyana had lower birth rates in comparison. The study estimated that 58718 children were born annually with a congenital heart defect in South America, with the highest incidences found in Brazil, Colombia, and Argentina, followed by Venezuela, Peru, Ecuador, Chile, Bolivia, Paraguay, Uruguay, and Guyana.

When examining the surgical treatment of congenital heart defects across South America, it becomes evident that certain countries display varying levels of success in providing adequate care. Specifically, Argentina, Chile, and Uruguay have established a commendable track record in terms of ensuring a high proportion of patients receive the necessary surgical procedures. These countries perform over 70 surgeries per million inhabitants, indicating a relatively strong capacity to diagnose and treat patients with congenital heart defects. Consequently, individuals from neighboring countries, such as Paraguay and Bolivia, often seek medical attention in the well-equipped hospitals of Argentina due to their robust public health system and expertise in the field.

On the other hand, countries like Peru, Venezuela, Ecuador, Bolivia, and Paraguay face significant challenges in bridging the gap between those in need of surgical intervention and those who actually receive it. These nations perform only around 20 surgeries per million inhabitants, indicating a substantial disparity between demand and access to care. Consequently, many families from these countries are compelled to migrate in search of the necessary medical attention for their children.

Brazil and Colombia fall somewhere in the middle of this spectrum, primarily due to the geographical features of their territories, which include regions with limited accessibility. Despite this challenge, efforts are being made to address the needs of congenital heart defect patients in these countries, albeit with varying degrees of success.

It has been reported that almost two-thirds of congenital heart defect patients require medical or surgical treatment within the first year of life, leading to approximately 41,000 new children

requiring such treatment each year in South America. Furthermore, nearly 40% of children who survived surgery would require one or two additional procedures for treatment completion, such as single-ventricle palliation or correction of late complications. This highlighted the potential increase in congenital cardiac procedures if the number of operations and survivors continued to rise. The study by Kreutzer and collaborators acknowledged improvements in healthcare policies regarding congenital heart defect coverage by 2011, particularly in the southern hemisphere¹. For instance, Chile and Uruguay showed excellent coverage, with 80% to 90% of children in need of congenital heart disease surgery receiving treatment. Argentina also witnessed significant coverage, close to 80%, thanks to a program supported by the Ministry of Health. It was believed that achieving full coverage for all patients could potentially lead to a two to three-point decrease in Argentina's infant mortality rate.

By collaborating with surgeons from each country, the researchers were also able to calculate the number of congenital cardiac procedures performed in South America. They found that over 17000 surgical procedures were conducted annually across the region, with more than 41000 new children requiring initial procedures for CHD treatment. At that time, there were 138 centers performing cardiac surgery, and more than 200 surgeons were involved in these procedures, although they were not exclusively dedicated to congenital heart diseases. On average, there was one center per 2.9 million people, and approximately 42 surgeries per million people were performed. However, the survey also revealed that 24081 children with a new congenital heart defect diagnosis each year did not receive any treatment. This highlighted a significant continental deficit of 58% (ranging from 12% to 86%) due to limited opportunities or the economic situation of individual countries.

By 2023, South America has experienced a considerable population growth of 15%, resulting in more than 8 million children being born in the region annually. Among these births, approximately 80,000 children are diagnosed with a congenital heart defect (CHD) each year, and out

of those cases, 40,000 children require surgical intervention. To address the increasing demand for specialized care, efforts have been made to train surgeons in renowned centers of the region or even in North America. These doctors return to their home countries, bringing their expertise and knowledge back to the region.

Recognizing the importance of improving congenital heart disease care, governments in South America have been enacting laws to mandate the creation and support of strategies to enhance the quality of care for individuals with congenital heart defects. These measures are intended to prioritize the development and improvement of programs specifically tailored to organize and streamline congenital heart disease care, ensuring that patients receive timely and appropriate treatment. These programs focus on aspects such as early diagnosis, accessibility to specialized centers, and the coordination of multidisciplinary teams.

As a result of these concerted efforts, some regions in South America have witnessed a decrease in the percentage of undiagnosed and untreated patients with congenital heart defects. This improvement can be attributed to enhanced awareness, increased availability of diagnostic tools, and improved referral systems, which facilitate the identification and timely intervention of affected individuals. Consequently, previously undiagnosed and untreated patients now have better access to the necessary healthcare services.

Moreover, the collective endeavors to improve congenital heart disease care in South America have yielded positive outcomes in mortality and morbidity rates. In certain regions, the implementation of these strategies and the consolidation of specialized centers have led to improved patient outcomes, reduced mortality rates, and decreased morbidity associated with congenital heart defects. In 2022, Argentina witnessed a significant improvement in healthcare outcomes as the overall mortality rate for the 3300 operations performed in the public sector stood at just 4%. These encouraging results serve as a testament to the effectiveness of the evolving healthcare landscape in the region and highlight the

potential for further advancements in congenital heart disease care throughout South America.

The paradigm of congenital heart disease care in South America

The paradigm of congenital heart disease care in South America presents a dichotomy. On the one hand, it is disheartening to acknowledge that only half of the patients in the region receive the necessary attention and care for their CHD. This lack of access to adequate healthcare resources can lead to adverse outcomes and reduced quality of life for many individuals affected by congenital heart defects. However, amidst these challenges, South America stands as the home of renowned surgeons who have made significant contributions to the field of pediatric and congenital heart surgery. These visionary surgeons have defied limited resources and infrastructure, striving to provide the best possible care to their patients. Their dedication, skill, and perseverance have led to groundbreaking advancements, establishing a rich history of achievements in the treatment of congenital heart diseases. Their contributions have not only impacted the lives of their patients but have also contributed to the global knowledge and understanding of congenital heart disease care.

One of the pioneers in our region was Euclides de Jesus Zerbini, a renowned Brazilian cardiac surgeon who has left an indelible mark on the field of cardiovascular surgery. His significant contributions have advanced the understanding and treatment of various cardiac conditions, particularly in the realm of congenital heart defects. One notable accomplishment of Zerbini is his groundbreaking study on the Tetralogy of Fallot, published in the *Journal of Thoracic and Cardiovascular Surgery* in 1965². This study, involving a large cohort of patients, showcased Zerbini's innovative approach to the total correction of the disease. The immediate results of this technique provided a new understanding of surgical interventions for this complex condition. In another significant publication in the *Journal of Thoracic and Cardiovascular Surgery* in 1969, Zerbini presented the long-term outcomes of these interventions, shedding light on the effectiveness and durability of the surgical treatment

employed and offering valuable insights into postoperative management and patient care³. But Zerbini's expertise extended beyond congenital heart defects. He delved into the realm of heart transplantation, contributing to the growing body of knowledge surrounding this revolutionary surgical procedure. In recognition of his remarkable contributions, Zerbini became the first South American member of the American Association for Thoracic Surgery and was bestowed with honorary membership in 1969, highlighting his international recognition and esteem within the field.

Adib Domingos Jatene was another renowned Brazilian cardiac surgeon who made significant contributions to cardiovascular surgery. His most notable achievement was the development of the Jatene operation -the proper name this technique should have- in 1975. Also called "the arterial switch operation", this groundbreaking procedure is used to correct the transposition of the great arteries in infants and has become the standard treatment for this complex congenital heart defect worldwide^{4,5}. Jatene's innovative surgical approaches have also advanced the repair of other complex congenital heart conditions, such as tetralogy of Fallot and truncus arteriosus, improving long-term outcomes for patients. In addition to his surgical expertise, Jatene played a pivotal role in shaping healthcare policies in Brazil. As Minister of Health, he implemented reforms that enhanced accessibility and quality of care, including the establishment of the Unified Health System. His advocacy for healthcare reform and universal access to medical services has had a profound impact on the Brazilian healthcare system, benefiting countless individuals. Throughout his career, Jatene has received numerous accolades and honors for his contributions to medicine and cardiac surgery. His innovative techniques, exceptional surgical skills, and dedication to patient welfare have solidified his reputation as a visionary leader in the field.

Guillermo Kreutzer, an Argentinian surgeon, now retired, is a highly regarded figure in the field of cardiac surgery. He has made significant contributions to the advancement of cardiovascular medicine. Notably, he is credited with co-inventing the Fontan-Kreutzer operation, a surgi-

cal procedure that has had a profound impact on the treatment of patients suffering from single ventricle⁶. His contributions were particularly crucial in certain concepts^{7,8}. He, along with his team, was the first to understand that atriopulmonary anastomosis would simply function as a pathway for the blood coming from the superior and inferior vena cava directly to the pulmonary circuit. Dr. Fontan believed that the right atrium would acquire the functions of the right ventricle and propel blood to the lungs and then to the left ventricle. Therefore, he placed valves both at the entrance to the right atrium and at its outlet. Dr. Kreuzer provided us with the understanding that in this type of univentricular palliation, the left ventricle serves as the aspirating pump, and therefore, valves not only do not serve a purpose but can even be detrimental by causing an obstruction. It was also Dr. Kreuzer who developed the concept of the fenestration and implemented it in all cases from his first patient onwards, arguing that it would allow for maintaining cardiac output at the expense of systemic cyanosis in the context of increased pulmonary resistance.

More recently, in 1993, Jose Pedro da Silva, another Brazilian innovator in the field of congenital heart surgery, invented and performed for the first time the cone operation, a technique used to reconstruct the tricuspid valve in patients with Ebstein's anomaly. This method aims to improve valve function, reduce tricuspid regurgitation, and enhance the overall quality of life for individuals with this anomaly, becoming the procedure of choice worldwide⁹.

While acknowledging the challenges faced in South America's CHD care landscape, it is crucial to recognize and celebrate the achievements and contributions of these remarkable surgeons. Their dedication and accomplishments serve as a source of inspiration and motivation for further progress in improving access to quality care for all individuals affected by CHD in the region. Innovation continues to thrive in the field of congenital heart surgery in South America, and our team has proudly contributed to these advancements. One notable achievement occurred in 2012 when we introduced a novel technique for repairing scimitar syndrome¹⁰. Our approach involves using the in situ pericardium as a conduit to redirect

the venous return of the right lung to the left atrium. By anastomosing the vein to the pericardium and creating a large conduit using the patient's own living tissue, we create a wide unobstructed tunnel that connects to a large opening in the lateral aspect of the left atrium. Remarkably, this procedure, known as the Lugones operation, has shown no incidence of postoperative obstruction after a decade of implementation, which contrasts with the high incidence of thrombosis associated with other techniques such as the baffle technique or direct reimplantation. As a result, the Lugones operation has been widely adopted by cardiac centers worldwide^{11,12,13,14,15}.

Additionally, our team has made significant contributions in the realm of semilunar valve reconstruction. These conditions, which can affect the aortic or pulmonary valve, pose considerable challenges in treatment. To address this, we have developed a method and device specifically for aortic valve reconstruction in pediatric patients. This innovative approach enables the construction of a new symmetric, oversized valve using the patient's tissue, allowing for growth accommodation. Rigorous testing in vitro and in vivo has yielded promising results, demonstrating excellent valve functionality^{16,17}. Currently, this strategy is being applied in clinical settings with encouraging outcomes.

In this dynamic region, innovation thrives as a prevailing norm. The scarcity of resources acts as a catalyst for minds to devise novel approaches, while the inherent flexibility of the system allows for experimentation and testing. However, it is important to acknowledge the profound disparity that exists, preventing a large segment of the population from accessing these advancements and improving their quality of life. To bridge this gap, it requires a collective effort from governments, private entities within the healthcare system, healthcare professionals, and society as a whole. Only through united action can we transform this reality and provide health and hope to millions of individuals across South America.

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Comprehensive program development for congenital heart disease in the United Arab Emirates and Singapore

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Abstract

Background/Objective: Establishment and development of tertiary-care centers for congenital heart disease (CHD) face multiple challenges as they need to be accessible, affordable/sustainable and provide the best possible outcomes. We report our experience of foundation a new tertiary-care CHD center in the United Arab Emirates (UAE) and program development in Singapore.

Methods: The prevalence and incidence of CHD in the context of local sociocultural environment was reviewed. Having established program elements of team, premises, and workflow, the quality assurance system was established and maintained by outcome analysis, and continuous audit in comparison with international databases.

Results: Consanguinity, segregated population pattern, and low prenatal screening rate increased the incidence/prevalence and complexity of CHD in the UAE. Increasing patient-load, strict professional working environment, strong governmental financial mandate for care appointed the centre into a market-leading position in the UAE. From the program start to 2020, we performed 4655 congenital cardiac operations with a median complexity=7.99 (v. international database mean=7; scale: 3-15) and hospital mortality=2.73±0.79% (v. expected mortality=3.41%). Singapore has a highly-developed, regulated, still competitive healthcare infrastructure, but low birth-rate and low prevalence of CHD represent challenges for program development started in 2021. Preliminary results of Singapore program development demonstrate improved team coherence, 60% growth of caseload, increasing case-complexity (from median=6.9 to 8.7), introduction of minimally invasive techniques (>80%) at preserving optimal outcomes (hospital mortality=2.5%). Additional CHD caseload from underserved regions could contribute to team performance maintenance to ensure the best possible patient and program outcomes in Singapore.

Conclusion: New and/or developing CHD centers face different challenges depending on the sociocultural and economic environment they function in. External demographic factors, case-load, complexity, referral patterns, healthcare financing structure are difficult to adjust; internal factors: program structure, quality assurance, professional working framework are subjects for improvement.

Introduction

New and developing centers for congenital heart disease (CHD) encounter a triple challenge as they aim to be (1) accessible to all ages and complexities and social backgrounds; (2) financially affordable/sustainable; and (3) they are expected to produce the best possible outcomes. This triple aim of modern healthcare is configured by external and internal factors. Demographic patterns, CHD prevalence/incidence, primary diagnosis framework and referral patterns presenting as public health demand are external factors that translate into case-load and complexity. Healthcare financing structure - acting as external/internal factor simultaneously - both regulates affordability for patients and families; and institutional sustainability. Internal factors, e.g., the characteristics of the actual multidisciplinary team, local quality assurance system and professional working framework affect the program structure¹.

The triple aim of accessible, affordable and high-quality congenital cardiac services is difficult to achieve in the real world that presents with global inequality of care^{2,3} and significant CHD populations are underserved worldwide⁴. Sending patients abroad for treatment is expensive, represents a temporary and individual solution, non-available for all; or it is simply impractical/impossible for time and/or geographical constraints. The establishment and development of services locally have been endeavored in the form of charity missions, cooperation/coaching programs, and foundation by resident international teams (**Table 1**).

Objective

We report two case scenarios for comprehen-

sive CHD programs by resident teams: (1) foundation of a new tertiary-care CHD center in the United Arab Emirates (Sheikh Khalida Medical City, Abu Dhabi; SKMC, UAE) and, (2) program development in Singapore (National University Heart Centre, NUHCS, Department of Cardiac, Vascular and Thoracic Surgery).

Materials and methods

Demographic patterns and prevalence/incidence of congenital heart disease was studied in national/international information sources^{5,6} to estimate CHD public health demand. In the program establishment scenario (SKMC), a local pediatric cardiology professional network was created first to establish primary diagnosis framework and referral patterns and follow-up pathways. As part of a feasibility study, healthcare financing structure was explored. An Institutional and organizational setting was established. Program preparation addressed facilities, equipment and the team from the viewpoint of continuum-of-care, expected caseload and case mix. Various segments of care, e.g., dedicated operating room, intensive care unit (ICU), high-dependency unit (HDU) and paediatric ward were assigned and equipped. Meantime, the international multidisciplinary team was recruited.

In both program establishment (SKMC) and development (NUHCS) scenarios, creation a professional working framework, rules of strict daily routines, multidisciplinary cooperation with team-empowerment, a robust quality assurance system including key performance indicators were agreed in accordance to international recommendations for congenital cardiac centers^{7,8}. We also employed various strategic, tactical and operational models to develop the specifics of a comprehensive service for CHD. Strategic models e.g., product/market grid, core compe-

Table 1: Advantages and disadvantages of different formats of CHD program development

Format	Benefits	Disadvantages
Visiting team missions	Individual patient benefit	No continuity-of-care Surgical safari Service disruption
Cooperation programs	Continuous external coaching Local talent advancement High professional standards	Brain-drain of local talent Applicability of adopted modalities
Resident teams	Continuity-of-care Local talent recruitment Organic program development	Sensitive financial sustainability Dependence on institutional support Possible slow program development

tencies, scenario-planning, etc. were employed in the planning/designing phase⁹. In the production phase, tactical models were added: e.g., 7-S framework, learning-v-traditional organizations for improving team dynamics¹⁰, etc.; and, the strategic model of strengths-weaknesses-opportunities-threats (SWOT) analysis⁹. Operational models, like root-cause-analysis served as continuous audit in form of a plan-do-check-act cycle¹¹. Quality assurance was maintained through weekly multidisciplinary team conferences, performance reviews, journal clubs. Clinical outcomes were measured with those in international databases^{12,13}. At SKMC program establishment, a special dispensation was sought with the health government to ensure the financial sustainability of the newly established CHD program.

Results

1. Establishment of a comprehensive service for CHD in the UAE

Historically, individual Emirati CHD patients were sent abroad for surgery/treatment; and no dedicated CHD service existed for resident non-nationals in the UAE. Our comprehensive program was established at Sheikh Khalifa Medical City (Abu Dhabi, UAE), a 588-bed, JCI-accredited government teaching hospital¹⁴. This flagship facility of UAE healthcare already hosted numerous tertiary-care surgical services: e.g., solid-organ transplantation, spine surgery, neurosurgery, etc. Our new, tertiary-care congenital cardiac program integrated into a cardiac institute, next to adult cardiology/cardiac surgery and a paediatric department already in existence.

Preliminary market research showed significant public health demand for CHD treatment and a medium-size, nationwide tertiary-care center was envisioned. A referral/admission protocol was circulated among a network of pediatric cardiologists and possible referring hospitals informing 24/7 availability of services. On the receiving side, in-house paediatric cardiologists were put in charge who communicated with critical care physicians and surgeons. During a 4-month-long preparation period a continuum-of-care protocol was drawn up for four complexity-based expected patient pathways: simplest (closed procedures), simple (e.g., ASD, VSD repairs), com-

plex (e.g., tetralogy of Fallot-repair, bidirectional/total cavopulmonary connection), most complex (neonatal open-heart procedures, e.g., arterial switch, Norwood procedure). These pathways prompted contingency aspects of the program e.g., dedicated OR, integrated pediatric cardiac ICU/HDU (9+6 beds), and service modalities that encompassed non-invasive/invasive diagnostics, catheter-based interventional, hybrid and surgical arms along with ECMO/ECLS. The pediatric cardiac multidisciplinary resident team comprised of 88 team members of 28 nationalities, speaking 39 languages that became a very cohesive resident group with no fluctuation of key medical professionals.

At the time of the founding of our CHD program, no comprehensive health insurance system existed in the UAE. Emirati nationals (15-18% of the population) were covered by statutory rights of their citizenship; however, residents (82-85%) lacked healthcare coverage. Pertinent UAE Health Law defined congenital heart disease as a *'life or limb emergency'* that allowed to admit *all* patients with CHD to SKMC. This mandate from the Health Authority Abu Dhabi ensured the financial sustainability of the service¹⁵. With the introduction of mandatory national insurance system (2011) and concomitant formation of various insurance companies, eligibility criteria for mandated care progressively stiffened and the government's financial involvement gradually decreased. (**Figure 1**)

Annual surgical volumes increased linearly in the formative years of the program (**Figure 2**). Subsequent plateau phase is attributed to a number of intrinsic and extrinsic factors. Institutional reorganization into separate paediatric cardiac surgery, anaesthesia, critical care, etc. divisions, rather than an integrated paediatric cardiac institute acted as an intrinsic barrier. Slowing intergovernmental projects reduced the influx of CHD patients from neighbouring countries and thus hampered SKMC's evolution into a regional center of excellence.

SKMC has been a sole provider for complex CHD in the UAE; that fact was represented in the distribution of age groups, complexity and acuity of the surgical procedures. Data divide into two

distinct cohorts: (1) neonates significantly higher complexity and acuity and (2) patients above 1 year of age. Survival, however, has not been significantly different between the groups (**Table 2**).

Higher representation of neonates and patients less than 1 year of age (71.1%) may ex-

plain the overall higher complexity in the surgical series. Comparison of historical local data on median complexity and hospital mortality rate with international dataset¹⁶ shows that local complexity exceeded the Database median value of 7.0 (Aristotle Basic Complexity Score: 3-15), whereas outcome represented by hospital mor-

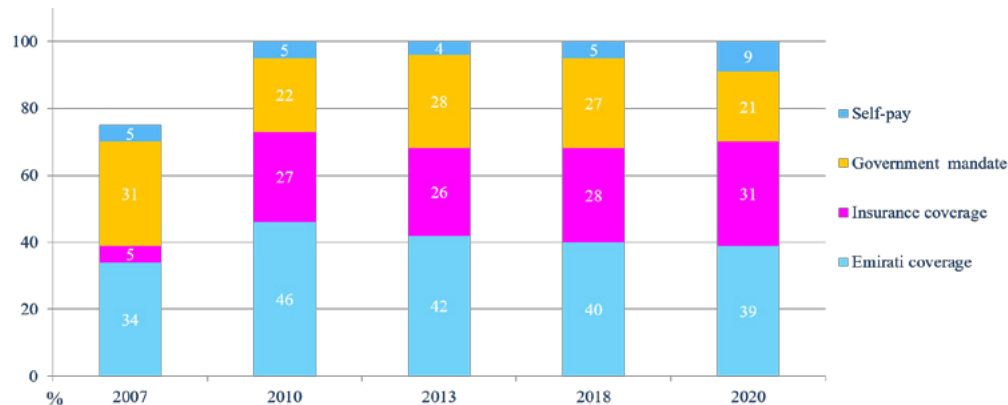


Figure 1. Financial coverage at SKMC Paediatric Cardiac Surgery, 2007-2020. Courtesy of Ms Huda Attiah

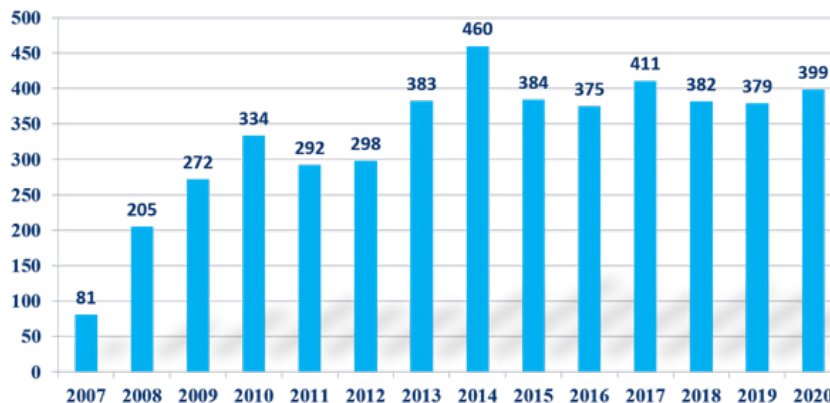


Figure 2. SKMC annual surgical volumes (2007-2020, N=4655)

Table 2. Distribution of age groups, complexity and acuity of CHD patients operated at SKMC (2007-2020, N=4655)

Age groups	Neonates	1month-1year	Beyond 1year	Statistical significance ^a
N=4655; primary procedures	1499 (32.2%)	1810 (38.9%)	1346 (28.9%)	Emergency neonates v electives beyond 1year
Elective	158	1091	1102	OR:366.08;
Urgent	1021	596	190	p-value:0.001
Emergency	320	123	54	
Complexity (mean, SD)	8.78 SD2.84	7.45 SD2.14	7.05 SD2.32	t-value:7.21381; p-value:0.00001
Survival (%)	96.92	97.93	98.24	OR:2.46; p-value:0.116; NS

'Elective' is defined as the surgical procedure is to be performed in the preferred time frame; 'urgent': within the same hospitalization; 'emergency': within 24-48 hours. Complexity is displayed on a continuous range of 1.5-15. ^aA two-tailed, paired Student's t-test was used for the comparison of numerical variables. P-value <0.05 was considered statistically significant. Categorical data were analysed by khi-square test and odds-ratio is provided (OR). NS: non-significant, SD: standard deviation

tality remained at par level of the international figures^{13,17} (**Figure 3**).

In want of a comprehensive service, numerous new modalities were introduced, e.g., 24/7 ECMO/ECLS service that opened for both perioperative cardiac and respiratory indications¹⁸. Similarly, hybrid techniques and intraoperative visualization, preoperative planning with 3D-printed patient-specific anatomic models were added¹⁹. Due to the sociodemographic dynamics of the UAE, Emirati patients were always overrepresented (34-46% in our surgical population v. 15-18% of Emiratis in the general popula-

tion). Adult congenital cardiac patients were still fewer and their gradual increase was expected once the patients locally operated on reached adolescent age²⁰.

A strengths - weaknesses - opportunities - threats (SWOT) analysis of the SKMC program (**Figure 4A**) reveals strengths as an established provider recognized by the community (both professional and public). A cohesive multidisciplinary team offers multimodality treatment for the entire spectrum of age and complexity. The weaknesses are related to the labor/cost intensive, staff/equipment sensitive high case-mix

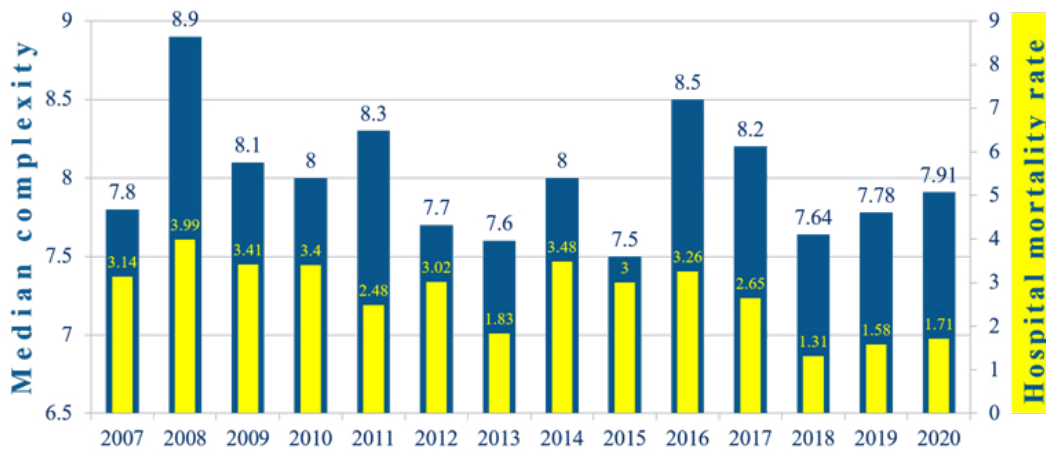


Figure 3. Median Complexity according to Aristotle Basic Complexity score and hospital mortality rates at SKMC Paediatric Cardiac Surgery, 2007-2020.



Figure 4. Comparative SWOT analysis. A: pediatric cardiac program establishment at SKMC, Abu Dhabi, UAE. B: program development at NUHCS, Singapore.

Abbreviations: CHD: congenital heart disease, COVID-19: coronavirus disease, CTVS: Department of Cardiac, Thoracic and Vascular Surgery, MDT: multidisciplinary team

index. Established treatment modalities, e.g., neonatal open-heart surgery, hybrid program, ACHD/GUCH-programme, ECMO, etc. create opportunities. Despite the service model is suitable for receiving trainees and it presents franchise opportunity (i.e., establishment of affiliated centers), the lack of local talent interest, and therefore, no incentive for education, poses a threat for long-term organic development.

2. Singapore program development

Singapore has a world-renown healthcare infrastructure and elaborated financing system that covers all her citizens and long-term residents²¹. Healthcare financing is governed by the principles of individual responsibility for health, patient co-payment to minimize moral hazard and wastage and government subsidies to keep basic healthcare affordable²².

The National University Heart Centre (NUHCS) - ranking 56th among the World's Best Specialized Hospitals²³ - is a tertiary referral hospital and academic medical centre affiliated with the National University of Singapore (NUS). The paediatric cardiac surgical program is part of the Department of Cardiac, Thoracic and Vascular Surgery (CTVS) which pioneers many innovative modalities, e.g., minimal invasive cardiothoracic surgery, hybrid and robotic programs, aortic surgery, etc. Paediatric and congenital cardiac surgery has a long tradition at NUHCS CTVS²⁴; however, it has remained a small program (there were 1081 surgeries performed for CHD between 2009-2020). A new vision for program development to become a provider of choice for complex CHD was initiated in 2021. Realizing that plan relies on 24/7 accessibility for emergency/urgent and elective CHD treatment. As treating CHD is a '*commitment for life*', the continuum-of-care spans from preoperative preparation throughout the intervention and postoperative period to the patient's journey back to and in the community. Family education, and home-monitoring is an integral part of that philosophy. Treatment is provided from neonatal complex repairs to adult congenital cardiac care (GUCH). A full spectrum of modalities is available, e.g., non-invasive/invasive diagnostics, catheter interventions, hybrid procedures, ECMO/ECLS, etc. Outcomes are measured at multidisciplinary team conferences,

and performance reviews with intraoperative video recordings on a weekly basis. Expected/observed progress and outcomes are also matched with international databases.

Initial (2021-2022) results of program development in Singapore do not allow a direct comparison with data from UAE, however, a comparison with institutional historical data²⁴ demonstrates a favorable trend, e.g., operative activity increased by 60% in the first two years - partially due to previous backlog; median case complexity also increased from median 6.9 to 8.7 (Aristotle Basic Complexity Score: 3-15) by internal audit. New procedures and modalities e.g., *en bloc* double-root rotation, hybrid approach (e.g., periventricular muscular VSD closure, intraoperative stenting, etc.) were introduced. Outcomes (hospital mortality=2.5%) are at par level with international database. Preoperative planning is supported by surgical emulation on 3D-printed prototypes and holograms. With cross-fertilization from a world-class minimally-invasive program at adult CTVS²⁵, this modality gradually finds its place in our pediatric practice. Currently, over 80% of the surgeries are performed as less-invasive procedures with minimal skin incision, with alternative perfusion techniques. Intraoperative video-recording of all procedures help performance improvement, training and education.

Training the next generation of surgeons is a key aspect of program development at NUHCS. Our team participates in postgraduate training schemes at both national (Postgraduate Medical Education, Specialist Accreditation Board of Singapore) and international levels (NUH International Training Fellowship). In a broader sense, we aim not just to treat but educate both professionals and the community.

A SWOT analysis of pediatric cardiac surgery work-in-progress at NUHCS is summarized on **Figure 4B**. NUHCS's multidisciplinary team has its strengths in its cohesion, open communication and excellent team-dynamics; however, the institutional organogram dictates that team members need to trespass their territorial boundaries to cooperate and improve quality of service. The team participates in charity mis-

sions (e.g., at Sri Satthya Sanjeevani Children's Heart Hospital, Suva, Fiji) where participants act out of their usual comfort zone in a high-volume environment; these avenues offer important opportunities for team-building and enhancing responsibilities. NUHCS pediatric cardiac services have a competitive edge of offering comprehensive services from pre/neonatal age to the grown-up congenital heart population by having a strong support from CTVS and its alliance with Khoo Teck Puat - National University Children's Medical Institute. Involvement into the planning for the new National University Hospital's infrastructure can reinforce mutual interdependence and contribute to a unique model of care. Affiliation with the National University of Singapore boosts research and education. Our current high case mix weakens optimal utilization of resources that could be ameliorated by reopening medical tourism. Singapore's low prevalence of CHD and consecutive smaller patient population - that threatens with suboptimal outcomes - mandates cooperation and networking among all involved entities at both institutional and national level. Concentrated action, i.e., cooperation seems the only viable option for nurturing local talent and training the next generation of professionals.

Discussion

Pediatric and congenital cardiac surgery is an essentially multidisciplinary discipline that re-

quires high investment, for being quantity-quality intensive, it poses a high threshold and faces high public expectations. **Table 3** illustrates both UAE and Singapore are high-income countries²⁶ that - theoretically - provides a strong financial basis for world class CHD care.

It is the local sociocultural context that a successful CHD service also needs to adjust and integrate into. The United Arab Emirates has a special social fabric as indigenous Emiratis (15-18% of the country's total population) and resident immigrants communities form separated population groups with little reproductive mixing²⁷. Consanguinity is traditionally and culturally prevalent²⁸. Previously high birth rate (16.5/1000 population; 2002) decreased (10,02/1000 population; 2022) to the level of the European average (10.42/1000 population; 2022)²⁹. Religious creeds would not allow termination of pregnancy³⁰. All these aspects contribute to an increased prevalence of CHD, and enhance their complexity³¹. Infant mortality rate radically decreased and has now reached to the range of the developed countries, signifying a rapidly advancing healthcare infrastructure³².

Singapore's cutting-edge healthcare infrastructure is organized in three clusters serving a population (5.98m; 2023) of Chinese (74.3%), Malay (13.5%), Indian (9%), and other (3.2%) subgroups with significant mixing and low consanguinity. Birth rate is at a low level and further

Table 3. Comparison of population demographics, economic data in the United Arab Emirates and Singapore in 2020²⁶. Estimates of CHD incidence and expected service provisions are based on the literature and international recommendations^{1,6}

	UAE	Singapore
Population	9.973.449	5.975.383
Median age (years)	38.4	35.6
Birth rate (1/1000 population)	10.76	8.94
Fertility rate	1.62	1.17
Infant mortality (1/1000 births)	5.06	1.54
GDP/capita (US dollars)	69.700	106.000
Healthcare expenditure (% of GDP)	5.5	6.1
Births	97.572	35.605
Neonates with CHD (1/170 live births=0.58% ¹)	566	207
Need for primary CHS (66% of CHD patients need surgery/year ⁶)	374	136
Staged/GUCH (10%/year ⁶)	38	14
Total CHS national need/year	412	150

Abbreviations: CHD: congenital heart disease, CHS: congenital heart surgery, GDP: gross domestic product, GUCH: grown-up congenital heart disease

decreasing (8.41/1000 population; 2022)³³. Prenatal discovery of CHD is about 95%³⁴; there is a general predilection for termination of complex anomalies (e.g., HLHS)³⁵. It is estimated around 200 neonates are born with CHD in Singapore annually. National demand for congenital cardiac surgery (even with staged and redo operations) does not meet the minimum output requirements of 250 operations for a tertiary congenital cardiac centre⁷. Singapore, a well-established hub for medical excellence had attracted medical tourism that somewhat compensated for the low caseload; however, influx of foreign patients completely halted during the COVID-pandemic. Increasing treatment costs also affect Singapore's competitive edge and hamper recovery of medical tourism.

Low caseload and case mix can lead to mediocre outcomes, a quantity-quality relationship often stated in congenital cardiac surgery^{7,36}, that prompts policymakers to centralize and professionals - ideally - to cooperate^{3,37}. Centralization and cooperation could become entangled in politics, institutional pride and pursuit of revenue³. Historically, congenital cardiac surgery is embedded in (1) children's hospitals where all the necessary allied disciplines are available; (2) conjoined to large adult cardiothoracic surgical departments where the paediatric subspecialty can rely on adult support services, supply-chain, etc.; or (3) - in case of major units - it functions as a stand-alone, dedicated congenital heart centre (of excellence) providing the full continuum-of-care. For a highly specialized discipline, congenital cardiac units are typically affiliated to teaching/university hospitals. Without denying the importance of the external and organizational factors, we propose that internal team dynamics, strict professional protocols and work ethics will ultimately decide the success of any congenital cardiac program. Team empowerment capitalizes on the professionalism, dedication and motivation of individual team-members and it acts for continuous improvement of a changing modality in a changing world (e.g., emergence of GUCH population, minimally-invasive hybrid modalities, etc.). Changing times also accentuate the moral responsibility for nurturing the next generation of professionals. Both program establishment (SKMC) and program development (NUHCS)

projects progressed in the form of *learning organizations v. traditional organizations*¹⁰. In learning organizations departmental boundaries are permeable in order to maximize skills to enhance creativity and learning and to ensure integrated processes as an approach to complex activities. A pediatric cardiac multidisciplinary team is the proper example of open, multifunctional networks among the divisions and departments and beyond.

Authors acknowledge the limitations of these two case presentations owing to personal experience and involvement in the respective programs that also bear the characteristics of real-time snapshots.

We conclude with corresponding quotes by Sheikh Zayed al Nahyan and Lee Kuan Yew founding fathers of the United Arab Emirates and Singapore that resonate the importance of education and handover to the next generation: *"The real asset of any advanced nation is its people, especially the educated ones, and the prosperity and success of the people are measured by the standard of their education."*³⁸ (Sheikh Zayed al Nahyan, 1918-2004). *"You begin your journey not knowing where it will take you. You have plans, you have dreams, but every now and again you have to take uncharted roads, face impassable mountains, cross treacherous rivers, be blocked by landslides and earthquakes. That's the way my life has been."*³⁹ (Lee Kuan Yew, 1923-2015).

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Nursing Education for Pediatric Cardiac Care Centers in Developing Countries

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Summary

Pediatric cardiac programs achieve excellent patient outcomes through the teamwork of a highly capable interdisciplinary team. Nurses are key members of this team and must possess a highly specialized set of skills and knowledge. A comprehensive nursing education program is needed to develop the necessary skills and knowledge to care for critically ill infants and children with heart disease who are often highly unstable, complex, and vulnerable. Orientation of novice nurses or those new to the unit begins with a needs assessment, resulting in a comprehensive plan to achieve the required competencies. Nurse residency programs are one method to provide a comprehensive orientation to novice nurses, using a combination of didactic instruction, precepted clinical experiences, and simulation. An ongoing nursing education plan is also important to refresh skills, especially with high-risk, low-volume procedures, as well as to meet regulatory requirements and develop new skills and knowledge. A key component of the pediatric cardiac nursing education plan is the development of preceptors, who provide hands-on instruction to mentor new nurses to develop the needed skills and knowledge while ensuring that patients receive safe care. Preceptor development can also provide experienced bedside nurses with opportunities to develop their knowledge and to continue professional development, which helps to positively influence job satisfaction. Numerous resources are available to develop a curriculum or the pediatric cardiac program. Evaluation is also an important component of the nursing education program to quantify individual as well as programmatic outcomes.

Introduction

Nursing care of infants and children with heart disease requires a highly specialized skills and knowledge to provide excellent care. The nurse's knowledge base needs to encompass the physiologic foundations of heart disease in infants and children, its treatment, and co-morbidities associated with heart disease. In addition, the nurse needs to have a broad understanding of psychosocial support, technical skills, leadership and critical thinking skills, teamwork, educational skills, and other areas. The purpose of this paper is to review the components and scope of nursing education specific to the care of pediatric cardiac patients and their families, as well as education evaluative strategies for pediatric cardiac programs in developing countries.

Characteristics of Critically Ill Pediatric Cardiac Patients

Infants and children affected by congenital heart disease (CHD) or acquired heart disease who require hospitalization and intervention require resource-intensive care. The bedside nurse must be highly skilled in the identification of early or subtle changes of cardiopulmonary instability, which may develop rapidly. In addition, the bedside nurse must be skilled at caring for patients that range in age from minutes after birth to early adulthood. The clinical manifestations of these patients may range from mild symptoms, such as tachypnea and poor feeding due to fatigue, to shock and cardiovascular collapse.

The American Association of Critical Care Nurses developed the Synergy Model for Patient Care in 1996 to conceptualize a nursing practice model, which is well suited to describe the needs of pediatric cardiac patients.¹ The primary assumption of the Synergy Model is that patient and family needs drive the required nursing competencies and skills; when the nurse competencies match the needs of the patient, unit, or system, synergy results and outcomes are optimized.¹ Patient characteristics span the wellness continuum to illness and include resiliency, vulnerability, stability, complexity, resource availability, participation in care and decision-making, and predictability.¹ For example, a neonate with

complex CHD and 22q11 microdeletion who underwent aortopulmonary shunt placement today may present as moderately resilient but highly vulnerable, unstable, complex, and unpredictable due to pulmonary over circulation and extracardiac effects of the genetic condition. The family may have few resources and require assistance to participate in care and decision-making. On the other hand, a five-year-old who underwent closure of a ventricular septal defect 24 hours previously is now on no vasoactive medications, and is breathing spontaneously is also moderately resilient but only moderately vulnerable and highly stable and minimally to moderately complex. Nurse competencies associated with the Synergy model are rated on a scale from one (competent) to five (expert) and include clinical judgement, advocacy and moral agency, caring practices, collaboration, systems thinking, response to diversity, facilitation of learning, and clinical inquiry.¹

Preparation for Practice

Nurses constitute the largest group of health-care workers globally.² Nursing preparation for clinical practice can vary widely from hospital-based to university-based schools or colleges of nursing. The World Health Organization defined outcome standards for nursing graduates at the entry into clinical practice level, including demonstrating established nursing competencies and understanding the determinants of health, meeting regulatory body standards for professional nursing licensure or registration, being awarded a professional degree, being eligible for entry to advanced education, and that schools employ tracking methods to quantify the success of graduates.³ It is important to recognize that nursing preparation for entry into clinical practice is at the generalist level. Pediatric-focused education and clinical experience are generally quite limited, and content related to the care of an infant or child with heart disease is even more limited, if present. In the South Pacific nation of Fiji, nurses are educated at one of two schools of nursing; one private and one university-based. Nursing graduates from these programs are overwhelmingly female (95%) and have a median age of 22 years.⁴

Impact of Nurses in the Pediatric Cardiac Intensive Care Unit

Intensive care, by nature, requires a highly specialized set of skills and knowledge implemented by a well-performing team of highly qualified, interdisciplinary healthcare professionals. A recent paper described the foundation of critical care as “Our machines and potions are impressive, but the most powerful ICU tool is--and likely always will be--skilled bedside staff, working in experienced teams, taught by experts, and supported by skilled colleagues.”⁵ Shortages of qualified and experienced nurses affected all countries prior to the COVID-19 pandemic, primarily due to an aging workforce and lack of nursing faculty.⁶ The COVID-19 pandemic exacerbated these issues and added new challenges, such as nurses leaving the profession to care for family members, occupational illness, growing levels of burnout and stress, and heavier workloads.⁶ Initial and ongoing specialty education then becomes a key factor to attract and retain highly qualified and engaged staff.

Although the entire interdisciplinary team is the vehicle through which excellent patient outcomes are realized, nurses play a uniquely important role. Bedside nurses with more than two years of experience are associated with decreased pediatric cardiac surgical mortality⁷ and decreased rate of cardiac arrest in pediatric cardiac patients.⁸ In addition, university-based pre-licensure preparation and specialty certification are associated with decreased complications in pediatric cardiac surgical patients.⁹

New Hire Orientation and Competency Assessment

The foundation of designing a unit orientation program, whether for nurses new to the nursing profession or experienced nurses new to the organization or unit, is a learning needs assessment.¹⁰ The needs assessment helps to develop a personalized educational plan that also helps the new nurse acquire the tools and knowledge to provide excellent care that meets the standards of care for infants and children with heart disease. A self-assessment of skills and knowledge is then followed up with skill validation or

checklists and targeted education.¹¹ Pediatric cardiac nursing orientation best practices include a variety of learning modalities with specific learning objectives and competencies, including a didactic curriculum, guided clinical experiences with a preceptor, and often simulation.^{12,13} Nurse residency programs utilize didactic instruction, precepted clinical experiences, role play, case studies, and simulation to facilitate the transition to clinical practice.¹⁴ In addition to unit-specific clinical content, the nurse residency program includes practice in delegation and prioritization, communication, conflict resolution, critical thinking, leadership, and socialization to the role. Nurse residency programs reduce novice nurse stress, build confidence, improve patient safety, and reduce nursing turnover rates.¹⁴ Nurses who have clinical experience but are new to the unit or organization also benefit from a comprehensive learning needs assessment to acknowledge the skills and experiences they bring with them, and to develop an organized learning plan to achieve unit competencies.

The curriculum for international pediatric cardiac nursing education has been suggested by the Pediatric Cardiac Intensive Care Society (PCICS) and is also available in a virtual platform (see **Table 1**).^{14,15} A useful method to divide up the neonatal and pediatric care content is to identify specific problems common to these groups, such as altered cardiac output, heart failure, pulmonary hypertension, cyanosis, malnutrition, single ventricle physiology, delayed neurodevelopment, altered family coping, and then use specific defects and diagnoses to illustrate these concepts. It is also important to include basic and advanced life support and recognition of clinical deterioration as core concepts.

Preceptors

Successful transition of bedside nurses to the pediatric cardiac care clinical setting requires a capable preceptor to guide the transition to practice. The preceptor has a vital role to ensure that safe patient care is maintained at all times while also guiding the new nurse to acquire new knowledge of unit and organizational policies and procedures, develop new psychomotor skills, and to create and implement an appropriate care plan.¹⁶

Table 1. Suggested Pediatric Cardiac Intensive Care Nursing Curriculum

Cardiac content	Professional practice content
Congenital cardiac defects	Precepting/mentoring
Neonatal cardiac care issues	Professionalism
Pediatric cardiac care issues	Staff support
Acquired heart disease	Moral distress and resilience
Arrhythmia management and pacing	Communication
Mechanical circulatory support	Supporting family coping
Nutrition	Team dynamics
Adult congenital heart disease	
Transition of care	
Pregnancy and heart disease	
Co-morbidities and other considerations	

Adapted from Zbylewski SC, Callow L, Beke DM, et al. Education and Training in Pediatric Cardiac Critical Care: International Perspectives. *World J Pediatr Congenit Heart Surg.* 2019;10(6):769-777. doi:10.1177/2150135119881369 and Virtual Nurse Curriculum Course <https://pcics.org/education/virtual-programs/>

Skilled preceptors are crucial and also help to socialize new nurses to their role and to the unit culture, as well as build the new nurse's confidence and knowledge. In the critical care setting, the preceptor helps the new nurse organize and prioritize in rapidly changing clinical situations, develop clinical reasoning skills, and respond effectively and empathetically to difficult situations such as death or end-of-life.¹⁶

An experienced and capable bedside nurse interested in becoming a preceptor should receive education on adult learning principles creating a learning plan, orientation expectations and competencies, evaluation strategies, effective communication and giving constructive feedback, and creative ways to work with a variety of learning styles.¹⁷ The preceptor should have support from an experienced nurse leader during the orientation process to problem solve challenging situations with orientees. Preceptors play an important role in transition to clinical practice and should receive commensurate recognition and opportunities for professional development.

Ongoing Education

Following unit orientation, it is imperative to develop an ongoing education plan for all nursing staff in the pediatric cardiac unit to maintain competency, provide professional development, and keep pace with evidence-based best practices.¹¹ This plan should encompass applicable regulatory requirements for ongoing skill validation, such as point-of-care testing or basic/advanced

life support. High-risk, low-volume clinical skills should be identified and a plan developed to ensure ongoing competency, as these skills such as renal replacement therapy, are infrequently needed but pose high patient risk because of their complexity.¹⁸ Some effective methods of ongoing skill validation include a Skills Fair day or skill stations set up in the unit, checklist-guided just-in-time education provided by unit clinical leaders, or online educational modules followed by knowledge and/or skill validation.^{11,18} The introduction of new equipment or procedures can also use these types of skill and knowledge validation.

Professional development opportunities, such as preceptor training, leadership development, and team building, may be more suited to a more didactic learning strategy, synchronous or asynchronous platforms. Didactic instruction could be made more engaging and meaningful using application strategies such as case studies or role play.

Simulation

Simulation is an exciting and intense educational strategy that can improve individual skills and knowledge, benefit team performance, patient safety, and decision-making capability.¹⁹ Simulation comes in many forms, from low-fidelity in-situ mock codes using a CPR manikin to specific skill trainers like a model arm to practice intravenous cannulation to high-fidelity complex simulation in a dedicated simulation lab. Low-fi-

delity simulation requires few resources, while a dedicated high-fidelity simulation laboratory may require a high level of resources to implement. Simulation provides training in affective communication skills as well as psychomotor skills, and can be adapted for an individual or for entire teams. Given that highly functioning teams are foundational to achieving excellent outcomes in the care of infants and children with heart disease, simulation should be considered an important component of nursing education, even if resources are limited to low-fidelity simulation.

Resources

As many wise people have said, trying to reinvent the wheel is foolish. Resources to develop a nursing curriculum for pediatric cardiac centers, in whole or in part, are readily available on the Internet, with zero to minimal cost. Examples of some of these resources are listed in **Table 2**. Other online learning opportunities may include university-based courses or specialty confer-

ences with remote attendance options; these generally require registration fees. The availability of virtual learning options has greatly expanded in the post-COVID pandemic era, and this type of learning can be cost-effective and high quality, especially if paired with follow-up application in a clinical setting.²⁰

Program Evaluation

The nursing educational program needs to be evaluated regularly, to quantify outcomes on an individual as well as program level. This important step is crucial to identify opportunities for improvement as well as needed revisions and updating. Different types of evaluation include formative, summative, process, outcome, and impact.²¹ Examples of each type of evaluation, specific to nursing education, are shown in **Table 3**. Evaluation of the nursing education program is vital to ease the transition to practice and improve nurse retention, which is especially important in this global shortage of skilled and

Table 2. Online Resources for Pediatric Cardiac Nursing Education

Website	Developer	Content Examples	Availability
https://www.heartuniversity.org/	Cincinnati Children's Hospital	Pediatric and adult CHD webinars, conference proceedings, grand rounds, lectures and classes, and guidelines.	Free, some content requires registration for a free user account
https://pcics.org/education/nursing-resources/	Pediatric Cardiac Intensive Care Society	Specific CHD lesions, postoperative care, nutrition, arrhythmias, pacemakers. Special interest groups are available to members. Virtual complete nursing curriculum available, but requires a fee.	Most resources are free. Individual or institutional membership fees based on country of residence.
https://learn.openpediatrics.org/learn	Boston Children's Hospital, Harvard Medical School	Extensive catalog of cardiovascular and critical care content covering assessment, specific types of CHD, postoperative care, management, procedures, hemodynamic monitoring, mechanical simulation, etc.	Registration required for free user account.

Table 3. Types of Evaluation in Nursing Education

Evaluation Type	Examples
Formative	Needs assessment (both individual and unit-wide), progress meetings and feedback, stakeholder analysis, SWOT (Strengths, weaknesses, opportunities, threats) analysis.
Summative	Satisfaction surveys, content examination scores, participant evaluation surveys, observations of patient care, patient/parent satisfaction surveys.
Process	Does the program meet its goals and objectives? Is it efficient? Are the learning methods feasible?
Outcome	Performance evaluations, completion of skills checklists and required competencies, chart audits, nurse-sensitive indicators (falls, pressure injuries, hospital-acquired infections).
Impact	Cost/benefit analysis, nursing retention, nurse vacancy rate, mortality, complications.

proficient frontline nurses.²² Nursing education programs also provide career development and advancement opportunities for preceptors and nurse educators, which may positively influence nurse retention.²³

Conclusions

Skilled and knowledgeable bedside nurses are the cornerstone of the interdisciplinary healthcare team and make significant contributions to excellent patient outcomes. Nursing education is the key to developing nurses as bedside experts and team contributors. Educational strategies begin with the transition to practice for novice nurses, continue with ongoing professional and career development, and incorporate multimodal, evidence-based learning strategies and evaluative methods.

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A post hoc analysis on the incidence of congenital heart disease in Baku-Azerbaijan calculated by a prospective epidemiology study

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Abstract

Background: The incidence of Congenital Heart Disease (CHD) in Azerbaijan was firstly published following a prospective study, using Echo-2D as a study method, in 2019.

Aim: Of this post hoc analysis of the first ever prospective epidemiology study that calculated incidence and types of CHD in Baku, Azerbaijan, is to verify the forms of diseases that will have an early clinical impact on the patients and the health system of the country.

Population-method: From June 2016 to August 2018, 2570, term neonates were screened in 2 major state maternity hospitals in Baku. Their screening was randomized to equal females/males with not known previous obstetric alert regarding CHD. Scanning was done by two teams of pediatric cardiologists by using echo-2D. Each team was 'blinded' to the findings of each other. All scans were recorded, and a third senior physician reevaluated them.

Results: From 2570 term neonates of the general population, they detected 47 CHD's. From them, 17 were critical and severe-CHD and 7/47(14.9%) were of moderate complex. 17/47 (36.2%) were cyanotic and 30/47(63.8%) were non-cyanotic. The incidence of simple CHD was 25/47(53.2%). Analysis of the specific anatomy is presented in table 1. The estimated incidence was 1.83%. As this incidence has been among the highest reported a post hoc analysis has clarified and presented an important clinical rate of 1.48% after redacting minimal defects without any clinical significance, as minor PDA's, ASD II, VSD's. A post hoc calculation of BAov, revealed an anatomical BAov incidence of 1.65% and a functional BAov incidence of 1.4%.

Conclusions: This first-ever prospective epidemiology study in Azerbaijan involving a cohort equal to 1.65% of the annual living births of the country, estimated a high incidence of CHD. This is among the highest reported globally. The amount of critical and severe CHD after the post hoc analysis increased from 46.8% to 55.3%. The incidence of cyanotic CHD after the post hoc analysis increased from 36.2% to 42.1%. These high numbers are possibly related to an isolated population and conjugated marriage customs of the country. As this represented a state population health burden a post hoc analysis based on clinical important CHD minimized the calculated index nearby 20%.

Introduction

Congenital Heart Diseases (CHD) are usually defined as clinically significant structural heart disease present at birth¹. The incidence of them in different studies varies from about 0.4% to 5% among the live births². Previous estimates of CHD came from few data sources, were geographically narrow, and did not evaluate CHD throughout the life course³. Additional to the lack of worldwide data, the method and age spectrum when CHD are detected play a critical role on the calculated incidence^{2,4}. For example, subaortic stenosis as well as valve lesions of Marfan's syndrome or obstruction due to hypertrophic cardiomyopathy and the clinical presentation of anatomical bicuspid valve almost always develops well after birth⁴. The use of echocardiography in the detection of CHD can "overestimate" the clinical importance of CHD. This can happen as rare and "self-treated" defects such as minimal alterations in the vena cava drainage, persistent left superior vena cava, draining in the coronary sinus or small muscular VSD's, ASD's II and silent PDA's can be included in an incidence or prevalence study of CHD⁴. These will not alternate the total burden of the disease as a state population health burden, so it will be better to exclude them from a national incidence study. These findings as well as the ability of the country to deal with a very sophisticated public health disease, lead or team to review our initial published data⁵ by a post hoc analysis and clarify a number that presents to the public health facilities of the country a population of patients that will need to be treated and followed-up.

Population - Method

In our initial paper⁵, we included a cohort of 2570 term neonates (delivered after the 38th week of gestation) that represented 1.65% of live births of the country, to increase the strength of our findings. During the period from June 2016 to August 2018, our cohort was screened in 2 major state maternity hospitals in Baku. Their screening was randomized to equal females/males, age four to five days old with not known previous obstetric alerts regarding CHD. Scanning was done by two teams of pediatric cardiologists – each team consisted by two experienced in

echo-2D pediatric cardiologists, using the same echo-2D device. This was a General Electric Vivid i cardiac ultrasound system, using an 8C-RS microconvex ultrasound transducer probe, suitable for neonatal high-quality scanning. Each team was "blinded" to the findings of each other team. All scans were recorded, and a third senior physician reevaluated them to determine the exact anatomy. When a conflict of opinion existed a majority of the five major researchers prevailed.

Results

From 2570 term neonates of the general population, they detected 47 CHD's. From them, 17 were cardiac CHD and 7/47(14.9%) were moderate complex CHD. 17/47 (36.2%) were cyanotic and 30/47(63.8%) were non-cyanotic. The incidence of simple CHD was 25/47(53.2%). Analysis of the specific anatomical types of CHD are presented in (**Table1**)⁵. The estimated incidence was 1.83%. As this incidence has been among the highest reported a post hoc analysis was undertaken aiming to clarify important clinical CHD that would receive a treatment in the first five years of life of the patient^{2,4}. Taking in consideration the natural history and outcomes of specific types of CHD, such as small ASDII's, small muscular VSD's and silent PDA's, we reviewed our data and excluded all not important clinical defects^{2,3,4}. So, 9 simple defects were reduced from the initial study. These represented 19.13% of the initial defects. Therefore, to that, the initial rate declined from 1.83% to 1.48 %. This represents a decline from the initial calculate incidence of 2019⁵ by 19.3%. The new analysis of the specific types of CHD are presented in (**Table2**) Also, we specified an updated incidence of BAov by a post hoc calculation that revealed an anatomic BAov incidence of 1.65% and a functional BAov incidence of 1.4%⁶.

Discussion

CHD's are a major cause of serious morbidity and mortality, among all age groups, worldwide; additional to that they are quite common and their - in many cases lifelong - management involves multidisciplinary teams of specialists leading to sophisticated services with a high cost^{3,4}. The incidence of congenital heart disease at birth

Table 1. Classification of CHD

1	BAov (functional/anatomic) no additional disease	3.035%
2	VSD (isolated/all types)	16.8%
3	ASD (isolated/all types)	8.4%
4	PDA (isolated)	6.3%
5	c-AVSD	4.2%
6	A-P window	2.1%
7	Coronary artery abnormalities	4.2%
8	PA valve stenosis	4.2%
9	Ao valve stenosis	4.2%
10	Mitral valve disease	2.1%
11	CoA (isolated/all types)	4.2%
12	IAA (isolated/all types)	2.1%
13	ToF (all types)	6.3%
14	d-TGA (all types)	4.2%
15	PAv atresia with IVS	4.2%
16	DORV (all types)	4.2%
17	TAPVD (all types)	4.2%
18	Tricuspid valve atresia	2.1%
19	HLHS	2.1%
20	c-c-TGA	2.1%
21	Truncus Arteriosus	2.1%
22	Ebstein Anomaly	2.1%
23	Univentricular Anatomy	2.1%
24	Heterotaxia Syndromes	2.1%
25	Shone's Complex	2.1%
26	Core triatriatum	2.1%

Table 2. Classification of CHD after post hoc analysis

1	BAov: [Functional 1.4%] & [True 1.65%] no additional disease	3.035%
2	VSD (isolated/all types)	7.9%
3	ASD (isolated/all types)	5.4%
4	PDA (isolated)	2.6%
5	c-AVSD	5.3%
6	A-P window	2.6%
7	Coronary artery abnormalities	2.6%
8	PA valve stenosis	5.3%
9	Ao valve stenosis	5.3%
10	Mitral valve disease	2.6%
11	CoA (isolated/all types)	5.3%
12	IAA (isolated/all types)	2.6%
13	ToF (all types)	7.9%
14	d-TGA (all types)	5.3%
15	PAv atresia with IVS	5.3%
16	DORV (all types)	5.3%
17	TAPVD (all types)	5.3%
18	Tricuspid valve atresia	2.6%
19	HLHS	2.6%
20	c-c-TGA	2.6%
21	Truncus Arteriosus	2.6%
22	Ebstein Anomaly	2.6%
23	Univentricular Anatomy	2.6%
24	Heterotaxia Syndromes	2.6%
25	Shone's Complex	2.6%
26	Core triatriatum	2.6%

(sometimes referred to as birth prevalence) depends on how a population is studied^{2,7}. Many incidence studies conclude now days to a figure close to 1.2% of live births. To this we need to add another 1.2% approximately of neonates that suffer from a BAov that at birth shows no pathology⁴. These seldom cause problems in childhood but account for many adult patients who require treatment for late-onset aortic stenosis or regurgitation. Any consideration of the burden of CHD must take these into account⁴. More, recent studies have shown a shift of the incidence of adults and median age of patients with severe CHD in the general population from the traditional pediatric age group, since 1985 to 2000. In 2000, there were nearly equal numbers of adults and children with severe CHD. This shift in figures is continues creating a larger adult than pediatric population of patients suffering from CHD⁸. So, this is the essence of CHD: a growing population worldwide of severe and critical forms of the diseases that will need a long-life

follow-up and multiple interventional and surgical treatment. On the level of State Health Services, we need to take in consideration that resources to treat CHD are both inadequate and seriously maldistributed worldwide⁴. The 2007–2009 World Society for Paediatric Heart Surgery Manpower Survey noted that about

75% of the world's population has no access to cardiac surgery, and that the distribution of cardiac surgeons as well as the was distribution of cardiovascular centers are very uneven, towards the needs of the bulk of the patients suffering from CHD⁹.

All the above-mentioned facts underline the need of an incidence that will be mostly orientated towards a clinical use than a rather academic one. These reasons led to the need of a post hoc analysis of our initial prospective epidemiology analysis, that still has a few weaknesses as it doesn't calculate a nationwide incidence of CHD and doesn't include data from fetal cardiol-

ogy that would represent a more accurate risk of CHD in the total population of the country.

Meanwhile the exception of tiny muscular VSD's with a diameter after birth of less than 1.0 mm,^{2,10,11}; ASD II's less than 5.0 mm^{2,12} and PDA's less than 1.0 mm in diameter^{2,13}, that can either spontaneously resolved or have none-specific clinical risk, amalgamates our calculated incidence to a more robust clinical index of 1.48 % regarding the exact index of CHD's that will need medical management.

In summary, the initial study⁵ showed a high incidence that has been reduced after our post hoc analysis, but still, a figure of 1.48% is higher than the mean worldwide reporter figure of 1.2%^{4,14}. The important message from the initial epidemiology study⁵ and its post hoc analysis is that when comparing to the standard western Europe -North Americas data both the critical and severe CHD as well as the percentage of the cyanotic forms of CHD are higher in Azerbaijan. Specifically, the average of critical and severe CHD in western Europe-North Americas is 25-30% and in both of our studies this number is between 46.8-55.3% and for the cyanotic forms of CHD from 20% in western Europe-North Americas, raised from 36.2-41.1%¹⁴. These data as well as the high incidence of functional BAov 1.65%, clearly indicate a specific spectrum of critical and severe forms of CHD, many of them cyanotic forms of CHD, that are not found in the standard epidemiology of western Europe -North Americas. Our personal explanation to these findings is a possibly related isolated population with a specific genotype in which a high-frequency conjugated marriage custom of the country owns this high and interesting phenotype of CHD's. These findings represent a state population health burden that will be addressed by specific policies in the nearby future.

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Mitral Valve Repair with a Biodegradable Annuloplasty Ring for Mitral Regurgitation

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Keywords

Biodegradable ring, annuloplasty, mitral regurgitation

Abstract

Objective: We aimed to evaluate the effectiveness of the mitral valve repair procedure using biodegradable ring implantation to treat severe mitral valve regurgitation.

Method: Between May 2007 and 2009, 20 patients without mitral stenosis underwent mitral valve repair using biodegradable annuloplasty ring. Preoperative, postoperative 7th day, 3rd month, 6th month, and 1st-year echocardiographic data of 20 patients were collected retrospectively and statistically analyzed with Chi-square and Student T-tests. 10- and 15-year results of 10 patients were available and the follow-ups were evaluated.

Results: At follow-up, transthoracic echocardiography revealed no or trivial regurgitation. Although there was an increase in the mean gradient during the early postoperative period (7th days and 3rd months), there was no statistically significant difference in the mean gradient between the preoperative and late postoperative period (6th month and 1st year). Mean gradients were 3.48 ± 0.64 , 2.72 ± 0.31 , 2.62 ± 0.25 , and 2.58 ± 0.21 mmHg, respectively at 7th days, 3rd months, 6th months, and 1st year. Increased preload due to mitral regurgitation resulted in larger left ventricular end-diastolic volume (LVEDV) and left ventricular end-systolic volume (LVESV) preoperatively of 194.42 ± 21.39 ml (range, 245.00 to 165.00 ml) and 94.23 ± 36.59 ml (range, 203.35 to 52.80 ml) respectively. The decrease in mean LVEDV and LVESV at 1 year was statistically significant, 130.40 ± 11.04 ml (range, 145.00 to 115.00 ml; $p < 0.001$) and 64.21 ± 13.20 ml (range, 80.08 to 40.25 ml; $p < 0.001$), respectively. Additionally, left ventricular ejection fraction (LVEF) reaches the preoperative value ($\%51.15 \pm 14.04$) in the 6th month ($\%51.58 \pm 7.32$), although there is a decline in the early postoperative period ($\%47.11 \pm 11.22$ on 7th day and 46.35 ± 8.03 in 3rd month). Long-term outcomes (10-15 years) were similar to those in the late-postoperative period (1st year). One late death occurred.

Conclusion: Annuloplasty using biodegradable mitral ring has shown desired late post-operative outcomes with particular advantages compared to traditional annuloplasty rings.

Introduction

Mitral valve repair is a more preferred method than prosthetic valve implantation in patient groups such as ischemic or degenerative mitral valve regurgitation, Barlow's disease, rheumatic origin mitral insufficiency, and congenital mitral valve disease. Ring annuloplasty during the surgical repair of mitral regurgitation significantly increases the success of valve repair.^{1,2} For this purpose, rigid and semi-rigid rings are used for ring annuloplasty. The recurrence of mitral regurgitation after implanting these rings was investigated in the long term.^{3,4} Studies have shown that the mitral annulus is not a fixed structure, and it shrinks with each ventricular systole, contributing to the left ventricular workload. This contribution was found to disappear with rigid ring implantation. Additionally, rigid ring annuloplasty to be performed in the pediatric patient group will not be able to follow the mitral annulus, which will expand with growing age. For this reason, deterioration in the mitral valve structure will occur. These problems were tried to be overcome with the method using biodegradable material.^{5,6} In this method, the aim is to create scarring in the mitral annulus and shrink the annulus after the inflammation thanks to the biodegradable ring. Since there is no rigid structure holding the annulus from the outside, it was predicted that the annulus would continue its physiological movements and would not prevent growth in the pediatric population. The first implantations of biodegradable rings in the pediatric population have had successful results^{7,8}. In the next period, the biodegradable ring was also used in the adult patient group.

Patients and Methods

In this study, the results of mitral annuloplasty operations using the biodegradable ring in adult patients with advanced mitral regurgitation were examined. Operations were performed at Maltepe University Cardiovascular Surgery Department between May 2007 and May 2009. Transthoracic and transesophageal echocardiography of all patients were performed by the same physician, using the Vivid 7 Dimension (General Electric Healthcare Systems, Milwaukee, WI, USA) echocardiography device in the

cardiology department. A transthoracic echocardiography examination was performed in the left lateral decubitus position by the guidelines of the American Society of Echocardiography. Left ventricular end-diastolic and end-systolic volume calculations were made in millimeters with the help of three-dimensional echocardiography, using the program installed in the echocardiography device. Mitral regurgitation was calculated with PISA (Proximal Iso Surface Area) method in the preoperative period. In this way, ERO (Effective Regurgitant Orifice area, cm²) and RV (Regurgitant Volume, milliliter) calculations were made. The calculations were repeated preoperatively and postoperatively (7th day, 3rd month, 6th month, 1, 10 and 15-years). The gradient formed on the mitral valve in the postoperative period was measured with CW Doppler (Continuous Wave Doppler) and the "mean" gradient was recorded. In addition, patient data in terms of tissue valve incompatibility, stenosis in the repaired valve, thrombus formation, pannus, rhythm changes in the heart, bleeding, infection, and stroke were investigated throughout clinical follow-up. The Chi-square test and Student-T test were used as statistical methods. $P < 0.05$ was considered statistically significant. The data in this study was analyzed with the SPSS 15.0 program. The mean age of the study group, which included 20 patients with mitral ring implantation, was 47.35 ± 13.13 . Seventeen of the patients were male and 3 were female. The (Body Mass Index) BMI of the patients was 25.89 ± 2.66 and their classification according to New York Heart Association (NYHA) was 2.3 ± 0.75 . The preoperative EuroSCORE values of the patients were 80% low risk (0-3 points), and 20% moderate risk (4-6 points).

Results

Two patients had ring implantation in both the mitral and tricuspid valves. As an additional surgical procedure; coronary bypass in 2 patients, chordal transfer in 4 patients, repair with Alfieri method in 1 patient, PFO/ASD closure in 1 patient, P2 chordae resection in 8 patients, and aortic valve homograft implantation in 1 patient were performed. The numbers of rings used were 30.45 ± 3.76 . Cross clamp time was 34.05 ± 9.62

minutes and cardiopulmonary bypass time was 51.25 ± 16.21 minutes. The amount of drainage from the patients was 472.50 ± 236.46 milliliters. The extubation time was 4.95 ± 2.23 hours. The length of stay in the intensive care unit was 1.70 ± 1.17 days.

Echocardiographic Evaluation Results

The main pathology in all patients included in the study was advanced mitral regurgitation. In the transthoracic echocardiographic examinations performed in the preoperative period, the ERO value was calculated as 0.44 ± 0.11 cm² and 54.42 ± 11.18 ml. In the postoperative pe-

riod, it regressed to the degree of trace or mild in the quantitative evaluation. While a moderate decrease was observed in the left ventricular ejection fraction values in the early postoperative (first 90 days) period, this decrease tended to improve in the later period. (**Figure 1, Table 1**). There was a statistically significant decrease between the LVEF values measured in the preoperative period and the early postoperative period (7th day, 3rd month) (Table 1). In the late postoperative period (6th month and 1st year) and long-term (10-15 years), there was a statistically significant increase in LVEF compared to the early postoperative period (**Table 1**).

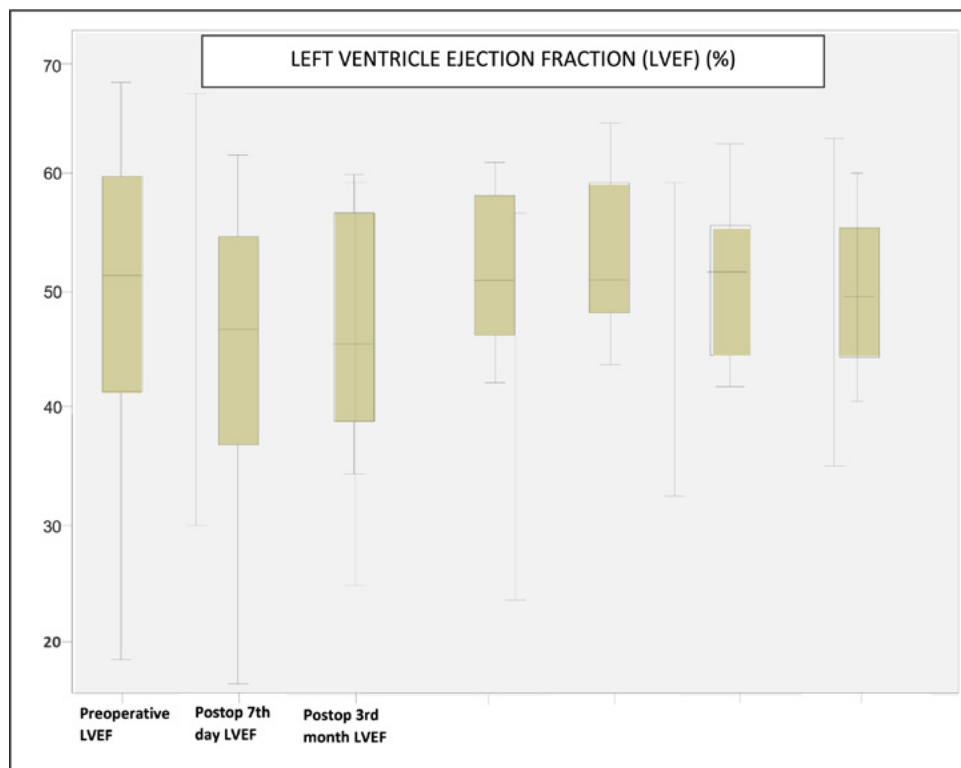


Figure 1. Left Ventricular Ejection Fraction Values as a Percentage

Table 1. Left Ventricle Ejection Fraction Values (Preoperative-Postoperative Period)

	N	Minimum	Maximum	MEAN	P value
PREOPERATIVE LV EJECTION FRACTION (%)	20	17	68	51,15 ± 14,04	<0,001
POSTOP LV EJECTION FRACTION 7th DAY (%)	19	16	62	47,11 ± 11,22	0,08
POSTOP LV EJECTION FRACTION 3rd MONTH (%)	17	34,00	60,00	46,35 ± 8,03	0,001
POSTOP LV EJECTION FRACTION 6th MONTH (%)	17	41,00	62,00	51,58 ± 7,32	0,10
POSTOP LV EJECTION FRACTION 1st YEAR (%)	10	43,00	65,00	51,10 ± 7,38	0,18
POSTOP LV EJECTION FRACTION 10th YEAR (%)	10	41,00	63,00	51,38 ± 7,32	0,05
POSTOP LV EJECTION FRACTION 15th YEAR (%)	10	40,00	60,00	49,36± 7,22	

In the postoperative follow-up of the patients, a significant decrease was observed in the left ventricular volume. This decrease was detected in both end-diastolic volume and end-systolic volume (**Figures 2 and 3, Table 2**).

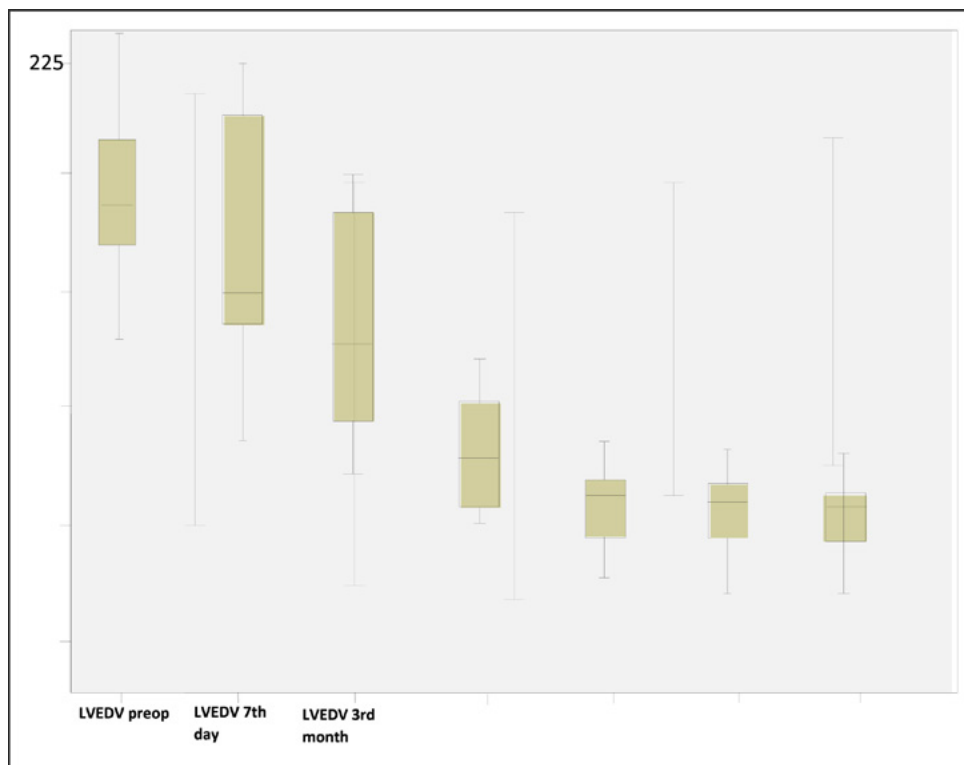


Figure 2. Preoperative and Postoperative Period Left Ventricle End-diastolic Volume (LVEDV) (milliliters)

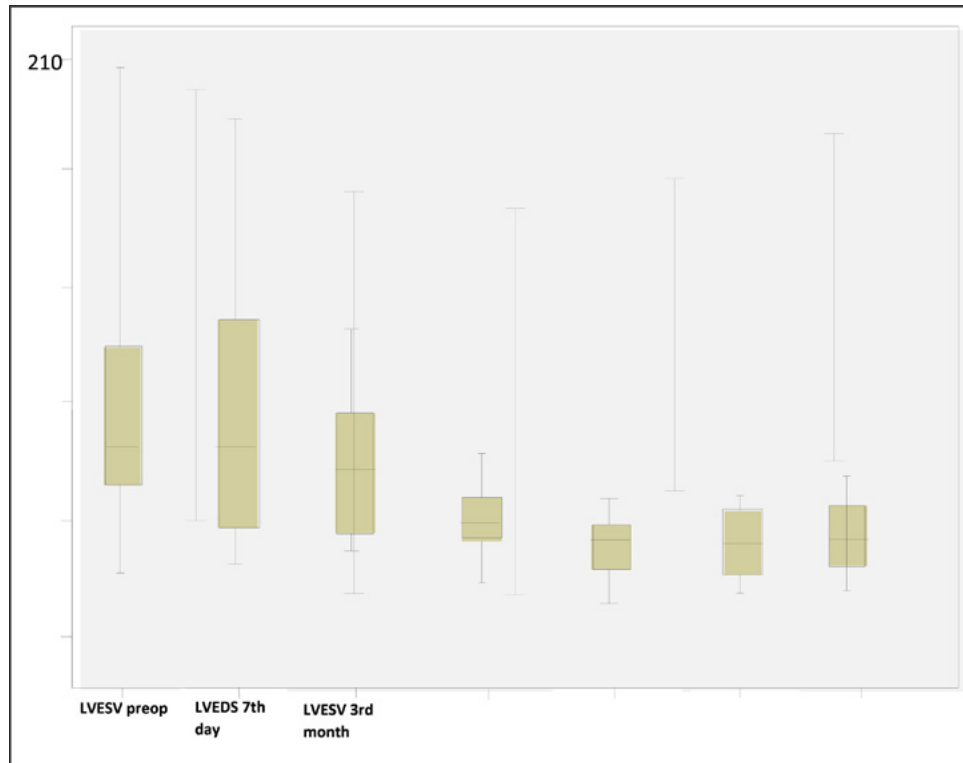


Figure 3. Preoperative and Postoperative Period Left Ventricle End-systolic Volume (LVESV) (milliliters)

Table 2. Left ventricle end-diastolic and end-systolic volumes

	Total N	Maximum	Minimum	Mean	Standard Deviation
LV ENDDIASTOLIC VOLUM PREOP	20	245,00	165,00	194,42	21,39
LV ENDDIASTOLIC VOLUM POSTOP 7th DAY	20	225,00	145,00	174,42	21,39
LV ENDDIASTOLIC VOLUM POSTOP 3rd MONTH	17	205,00	140,00	159,37	17,14
LV ENDDIASTOLIC VOLUM POSTOP 6th MONTH	17	160,00	125,00	141,18	10,60
LV ENDDIASTOLIC VOLUM POSTOP 1st YEAR	10	145,00	115,00	130,40	11,04
LV ENDDIASTOLIC VOLUM POSTOP 10th YEAR	10	142,00	110,00	128,00	12,07
LV ENDDIASTOLIC VOLUM POSTOP 15th YEAR	10	142,00	110,00	126,00	10,01
LV ENDSISTOLIC VOLUM PREOP	20	203,35	52,80	94,23	36,59
LV ENDSISTOLIC VOLUM POSTOP 7th DAY	20	189,00	56,55	94,34	32,67
LV ENDSISTOLIC VOLUM POSTOP 3rd MONTH	17	125,40	60,00	85,42	19,26
LV ENDSISTOLIC VOLUM POSTOP 6th MONTH	17	91,20	47,50	68,88	14,61
LV ENDSISTOLIC VOLUM POSTOP 1st YEAR	10	80,08	40,25	64,21	13,20
LV ENDSISTOLIC VOLUM POSTOP 10th YEAR	10	81,02	43,70	65,07	12,10
LV ENDSISTOLIC VOLUM POSTOP 15th YEAR	10	85,08	44,25	67,21	13,25

In the transthoracic echocardiography performed in the preoperative period, no gradient was detected on the mitral valve to indicate the formation of stenosis. However, transthoracic echocardiography performed in the early postoperative period revealed a mild to the moderate

gradient on the mitral valve. This gradient formation gradually decreased in the late postoperative period and returned to normal levels at the end of 1 year. There was no significant difference in 10- and 15-years. (Figure 4 and Table 3).

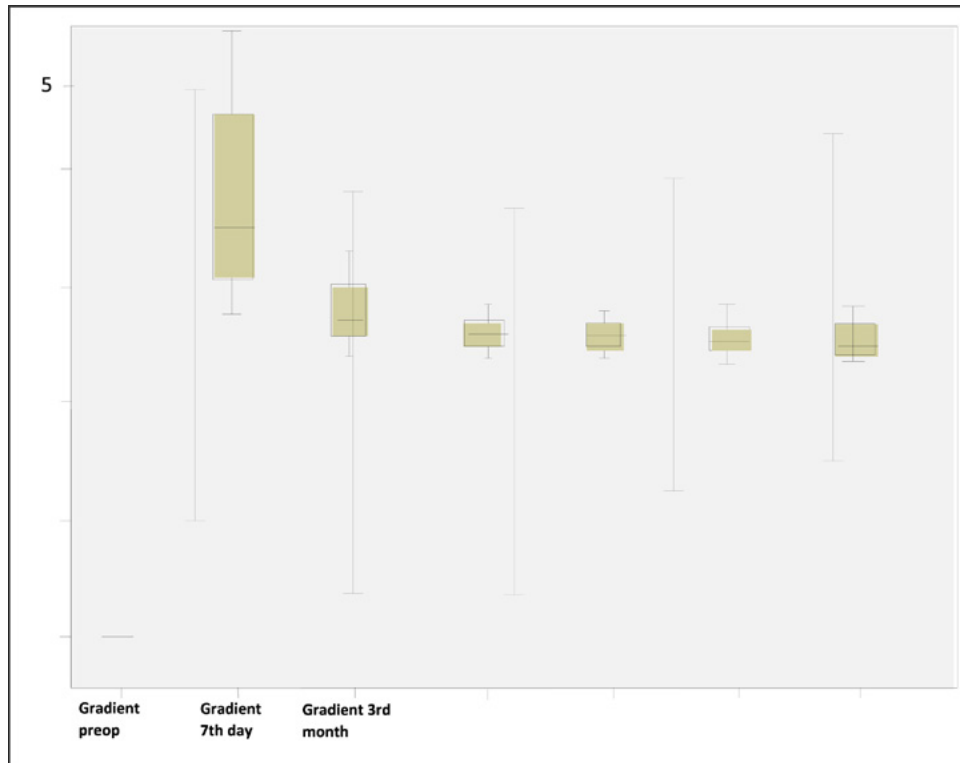


Figure 4. Gradient values on the mitral valve in the preoperative and postoperative period (mm Hg)

Table 3. Gradient values on the mitral valve in the preoperative and postoperative period

	Total N	Maximum	Minimum	Mean	Standard Deviation
PREOP GRADIENT on the VALVE (mmHg)	20	,00	,00	,00	,00
POSTOP 7th DAY GRADIENT on the VALVE (mmHg)	20	5,80	2,90	3,48	,64
POSTOP 3rd MONTH GRADIENT on the VALVE (mmHg)	17	3,20	2,40	2,72	,31
POSTOP 6th MONTH GRADIENT on the VALVE (mmHg)	17	2,90	2,30	2,62	,25
POSTOP 1st YEAR GRADIENT on the VALVE (mmHg)	10	2,80	2,30	2,58	,21
POSTOP 10th YEAR GRADIENT on the VALVE (mmHg)	10	2,90	2,30	2,50	,17
POSTOP 15th YEAR GRADIENT on the VALVE (mmHg)	10	2,95	2,35	2,42	,20

Mitral regurgitation greater than or equal to +2 developed during the follow-up period in 3 patients. Mitral regurgitation regressed in the serial ECHO controls of the 2 patients. The other patient was re-operated for mitral valve replacement. Late death occurred in 3 patients during the follow-up period. 2 of them were due to non-cardiac events, carcinoma. Cardiac-related death occurred in one patient due to complications caused by arrhythmia. During the follow-up periods, the 10-year and 15-year survival rate were 90% and 90%, respectively.

Discussion

Mitral valve repair techniques have shown

promising results with lower operative mortality, avoidance of anticoagulation, better preservation of left ventricular function, and the possibility of the continued growth of the valve in young children⁹. An annuloplasty ring has been used frequently for mitral valve repair since the late 1960s when the first generation human rigid mitral valve ring was introduced. The annuloplasty ring is sutured onto the native annulus to correct dilatation, consolidate the valve repair, improve leaflet coaptation during systole, and remodel the shape of the mitral valve. Traditional annuloplasty rings may cover the needs of the adult population, but it brings about suboptimal valve repair in pediatric patients. In addition, the classic annuloplasty ring used in the pediatric group has

two major drawbacks directly related to the ring. These drawbacks are exposure to foreign material causing a risk of fibrous tissue overgrowth, which may have an impact on valve function and restricted potential for native annular growth¹⁰.

Kalangos biodegradable annuloplasty ring (Bioring SA, Lonay, Switzerland), commercially available in sizes 16 to 36 mm. This biodegradable ring has a curved C-shaped segment of poly-1,4-dioxanone polymer colored with a blue dye, which makes it only a partial ring. The ring is attached at both ends with a needle-holding extension suture (2/0 monofilament polyvinyl). The ring is implanted into the posterior annulus using the suture extension and fixed at the anterior and posterior trigones of the mitral valve. It represents a new concept owing to its biodegradable properties and enables us to apply a unique intra-annular implantation technique. This ring encircles the entire length of the posterior segment of the mitral valve, as well as commissural areas, and supports the posterior annulus from trigone to trigone¹¹. Although the ring does not encircle the entire annulus, this can be considered acceptable because the results of posterior annuloplasty have been suggested to be equivalent to circumferential annuloplasty.

The semi-rigid structure of the ring enhances leaflet coaptation at the time of implantation and permits mobility of the mitral valve annulus during the cardiac cycle, yet it does not interfere with the motion of the posterior annulus and the leaflets. The gradual biodegradation of the ring induces annular fibrosis and ensures optimal annular reinforcement and satisfying results in the midterm. Although the concept of mitral-annular biodegradable ring implantation is novel and may raise concerns about the safety of its implantation technique, experimental studies^{12,13} have confirmed that Kalangos biodegradable annuloplasty ring does not cause ischemic complications secondary to circumflex coronary artery occlusion¹⁴. Kalangos biodegradable ring annuloplasty is facilitated by a single continuous suture, unlike the multiple interrupted sutures used in conventional annuloplasty. Hence the implantation time and the aortic cross-clamp time are significantly shorter¹¹. This is particularly important in complex mitral valve repairs and concom-

itant procedures^{15,16}. The intra-annular position of the ring prevents contact with the blood, thus, it negates the need for anticoagulation. Moreover, the ring preserves the growth potential of the native mitral and tricuspid annulus. Conventional annuloplasty rings consist of woven, non-degradable prosthetic material that may be a source for the proliferation and colonization of bacteria and may adversely affect the surgical outcome^{17,18}. The sub-endocardial implantation of the biodegradable ring prevents direct contact of the ring with the blood circulation; therefore the risk of postoperative infection decreases especially for patients with infective endocarditis. Pektok et al. have indicated this benefit in their study using biodegradable rings in 17 consecutive patients with acute infective endocarditis¹⁹.

Yakub et al. shared their experience regarding the implantation of the biodegradable ring in young children between 2006 and 2011. 68 patients underwent mitral valve repair for congenital mitral valve disease. They divided their patients into two groups, which were 39 patients with biodegradable annuloplasty ring implantation, and 29 patients with non-ring annuloplasty techniques. There was a significant difference between the two groups concerning freedom from mitral valve repair failure ($p=0.04$) and mitral valve re-operation free survival ($p=0.026$). Echocardiography follow-up on 24 patients with the biodegradable ring was undertaken to assess the growth of the native annulus. The mean Z-score was noted to undergo normalization at 3 and 5 years, which suggests normal annular growth. This is another advantage of the biodegradable ring in the pediatric population since it permits the growth of the native annulus²⁰.

We tried to evaluate the early and late results of hemodynamic changes in patients who underwent mitral valve repair with a biodegradable ring, keeping echocardiographic findings in the foreground. The usage areas and results of the biodegradable annuloplasty ring in different patient groups have been previously examined in many studies mentioned above.

With this study, we saw that the advanced degree of mitral regurgitation decreased to a trace or mild degree in the post-op period. Although

LVEF decreases in the early postoperative period (7th day and 3rd month), there is no statistically significant difference in LVEF between the pre-operative and late postoperative period (6th month and 1st year) and long-term (10- and 15-years) as well. This situation was interpreted as the sudden increase in afterload as a result of acute correction of mitral regurgitation. Another important finding was a significant decrease in left ventricular volumes in the post-operative follow-up. This decrease occurred in both end-diastolic volume and end-systolic volume. In addition, no gradient was detected on the mitral valve in the examinations performed with transthoracic echocardiography in the pre-operative period. However, there was a mild to the moderate gradient on the mitral valve in the early post-op period. This gradient formation gradually decreased towards the late post-op period. When the studies with rigid and semi-rigid rings were examined, the continuation of the gradient on the mitral valve during the late postoperative period was an important problem^{21,22,23,9}. Therefore, a biodegradable mitral annuloplasty ring might be a potential option to treat severe mitral valve insufficiency.

In the literature, there are studies showing that the long-term results are more satisfactory since the repair surgeries using flexible rings are more physiological repair method^{24,25}.

In our patient group, the main pathology was advanced mitral regurgitation. It was a male predominant group in the younger age group than the patient population examined in the studies in the literature. It was an interesting result that 2 patients with +2 or more mitral regurgitation in the early post-op period were free of mitral regurgitation in their long-term follow-up. The possible reason is that it is a physiological repair method and that the biodegradable ring interposed intra annularly allows fibrosis and ventricular remodeling.

Conclusion

The biodegradable ring can be implanted successfully to treat mitral regurgitation resulting from various etiologies. The more widespread use of this ring is expected with the disclosure of

long-term results of ongoing clinical studies.

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Conflict of Interest

Authors claim no conflict of interest

Study Limitations

An important limitation of this study is the small number of patients. This may cause to miss some statistically significant values. Depending on the number of cases, data significance that would allow the comparison of case groups such as ischemic and degenerative mitral regurgitation could not be achieved. In addition, the absence of a comparison group like the valve replacement patient group is another limitation.

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