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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 ontribute 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 America[1]. 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 700000 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 41000 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 hemisphere[1]. 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 1965[2]. 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 care[3]. 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 ventricle[6]. 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. Kreutzer 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. Kreutzer 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 worldwide[9].

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 syndrome[10]. 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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