



Cardiosurgical Care in Case of Congenital Heart Defects: Modern Approaches and Features of the State of Patients' Health

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Authors' contributions

This work was carried out in collaboration among all authors. Author LAE and ISG designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Authors DRM and TT managed the analyses of the study. Authors FAD and AAS managed the literature searches. All authors read and approved the final manuscript.

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Short Communication

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ABSTRACT

Congenital heart defects today, despite the early intrauterine diagnosis, are still quite common, and their danger to the patient's health is significant. The article discusses the features and modern approaches to the provision of cardiac surgical care for this congenital disease. The author comes to the conclusion that modern cardiac surgery technologies, rooted in the distant 50s of the twentieth century, allow us to conclude that at the present stage, surgical intervention in case of congenital heart defects has advanced far. Despite the challenges in this area, cardiac surgeons continue to make progress in reducing the mortality and multiple comorbidities of patients with congenital heart defects.

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1. INTRODUCTION

Congenital anomalies are defined as structural and functional disorders that develop during pregnancy and can be diagnosed prenatally, at birth, or in childhood.

This year, the World Health Organization highlighted the importance of early diagnosis of various heart defects in one of its reports, naming them as the cause of about 5.0% of neonatal deaths worldwide, as well as 8.0% of post-neonatal deaths. For example, approximately 3 million children are diagnosed with severe heart defects each year, and this number explains 303,000 newborns who die within four weeks of birth due to this type of abnormality [1].

The first operations for patients with a diagnosis of "heart disease" belong to the period of the 50s of the twentieth century, even then specialists made attempts to reduce the mortality of patients with this diagnosis and improve their quality of life.

The diagnosis and treatment of congenital heart disease, the most common birth defect, is a huge success story of modern medicine. In the 1950s, the survival rate of children born with CHD was only about 15%, whereas today more than 90% of these children live to adulthood.

In 1938, Robert E. Gross, at the Children's Hospital in Boston, performed the first successful operation to correct an extracardial lesion, namely, a ligation of the ductus arteriosus. This operation really represents the first milestone in the treatment of congenital heart defects [2].

Shortly thereafter, in 1944, Clarence Craford performed the first successful restoration of aortic coarctation at the Karolinska Hospital in Sweden. [3] In the same year, at Johns Hopkins Hospital in Baltimore, surgeon Alfred Blalock and pediatric cardiologist Helen B. Taussig developed a shunt to facilitate the treatment of patients with cyanotic congenital heart disease [4]. The Blalock-Taussig shunt, connecting the subclavian and pulmonary arteries, increased pulmonary blood flow in cyanotic infants with pulmonary stenosis, improving their well-being within minutes. Notably, Taussig later became the first woman and first pediatrician to be elected president of the American Heart Association.

In the 1950s, research on congenital heart defects was largely focused on surgery and various methods of applying hypothermia in clinical practice. In fact, the method of open-heart hypothermia was first used by F. John Lewis in 1952 on a human to close an atrial septal defect at the University of Minnesota [2]. However, hypothermia allowed operations to be performed only with simple defects, as their time was reduced by 5-10 minutes of safe stopping time, so as not to endanger the central nervous system. More complex lesions were still inaccessible to surgical intervention [2].

The following year, in 1953, John Gibbon used a cardiopulmonary bypass machine for the first time in history to repair an atrial septal defect, which provided additional protection for vital organs [5]. Although many surgeons knew about the Gibbon heart-lung machine, the first machines were huge and complex. What was needed was a simple and effective means of performing surgery on a bloodless, somewhat immobile heart, with sufficient time to correct defects that were more complex than those that could only be repaired by hypothermia.

After a series of experiments on dog models in 1954, C. Walton Lillehei performed the first case of restoring Fallot tetralogy on an 11-year-old boy using controlled cross-circulation. [3] The donor was a volunteer from the boy's hometown, as neither parent was compatible. The donor's femoral artery was connected directly to the recipient's right common carotid artery, and the recipient's external jugular vein was connected to the donor's femoral vein using plastic cannulas. One pump was used for both systemic arterial and venous blood flow. The cannula, which was inserted into the recipient's jugular vein, advanced to the inferior vena cava and had lateral openings that allowed venous blood to be diverted from both the inferior and superior vena cava.

Due to the fact that this procedure was new, Lillehei decided to limit its use and used it only for those patients whose survival without surgery was zero [6]. This was the only operation described with a potential fatality rate of 200%. A parent could lose not only their child, but also their spouse, who in most cases acted as a donor. Despite the initial perioperative mortality (four out of ten patients died), Lillehei followed

this path. His method paved the way for open-heart surgery, and Lillehei rightly known as the "father of open-heart surgery."

In the area of operations for congenital heart defects, many innovative procedures have been performed. In 1967, Donald Ross in London replaced the diseased aortic valve with the patient's own healthy pulmonary valve, placing the homograft in the pulmonary position. [2] This procedure is now commonly known as the Ross procedure. While this still causes problems, such as technical complexity and potential long-term failure of two valves, there is evidence to suggest that long-term outcomes in properly selected young patients, including women planning pregnancy, may be a better choice compared to traditional aortic valve replacement. [2] To date, the Ross procedure has been successfully applied.

Thus, from the 1950s to the present, there has been a dramatic increase in the total number of operations for congenital heart defects. Thanks to improved diagnostic techniques combined with advances in surgery, cardiology, and intensive care (including the use of prostaglandins, inotropes, nitric oxide, extracorporeal membrane oxygenators, and ventricular assist devices), operational mortality has significantly decreased, and long-term survival has improved over time [7].

As patients with congenital heart defects get older, their mortality rate increases compared to the general population, with the highest mortality rate among patients with complex congenital heart defects. Currently, the leading cause of death in adults with congenital heart defects is heart failure [3].

Current methods of cardiac surgery are constantly being upgraded, and this can be illustrated by a number of examples. In particular, methods of aortic valve surgery ensure the growth of the involved valve, low thrombogenicity and immunogenicity, and minimize the frequency of repeated surgical interventions, giving patients the highest chances of survival.

There is still debate about whether primary interventional balloon dilation or surgical treatment of congenital aortic stenosis will be more effective. At the same time, it has now become clear that in adolescents and young adults, replacing the valve with any other substitute, with the exception of a pulmonary

autograft, leads to a reduction in the life of patients by 20 years [4].

One example of progress in surgical technology is the improvement of the previously described techniques. One of the best examples is the Starnes procedure. At the turn of the century, the vast majority of newborns with Ebstein's disease died with severe symptoms, but now this procedure can reduce infant mortality.

Today, technologies are also used for improved fluid drainage during the connection of the patient to the artificial blood circulation. New cardioplegia regimens, such as single-cell depolarizing intracellular solutions, histidine-tryptophan-ketoglutarate solution, or del Nido solution, are also becoming widespread [6].

Currently, continuous detailed monitoring of patients in the intensive care unit for a variety of parameters is the standard of care in developed countries. Today, there are high hopes for building an analysis of all this data in real time using artificial intelligence to predict adverse events, in particular, cardiac arrest.

Over the past few decades, combined therapeutic strategies of interventional catheterization and surgical techniques have been used to treat patients with congenital heart defects better, mainly in the pediatric population [3].

Also, in cardiac surgery today, developments are used that were released at the end of the last century, but for a long time they did not receive practical application. In particular, the use of the "Berlin Heart", which was developed in the nineties, began to be used only at the turn of the century, but quickly became an important aspect of heart transplantation in children [6].

The development of small implantable assistive devices, such as Heartware, has made it possible to use them among adolescents. A portable implantable device that can be implanted in young children for long periods of time may soon be available, opening the door to its use in the most severe of our neonatal patients [3].

Tissue engineering, stem cells, regenerative technologies, and mitochondrial implantation are also used in cardiac surgery today. In particular, smooth muscle and endothelial cells, stem and

mononuclear cells, etc. are used to create artificial cardiac tissues.

The discovery of cardiac progenitor cells challenged the dogma that the heart is a post-mitotic organ and opened the door to cellular regenerative therapy for heart disease. Since then, several preclinical and clinical studies have been conducted on various types of pluripotent stem cells and different delivery methods in adult and pediatric patients with heart disease. The results of the first therapeutic attempts were disappointing for adults, but promising for some populations of children, especially with one ventricle. Initial results highlighted the safety of the procedure. Results in terms of the effectiveness of improving cardiac function are still expected.

Recently, there has been considerable interest in the new and developing science of mitochondrial transplantation [7].

The steps of mitochondrial transplantation are summarized as collecting viable tissues such as skeletal muscle, isolating mitochondria, testing a sample for viable mitochondria, and injecting extracellular mitochondria into risk tissue cells such as the myocardium. These mitochondria

undergo internalization and participate in cellular ATF generation, mitochondrial DNA repair, and cytoprotective cytokines.

Three-dimensional modeling and printing is another area in which the connection between a surgical scientist and engineers promises good prospects in the field of surgery for congenital heart defects. Three-dimensional modeling can be attributed to the virtual three-dimensional reconstruction of the patient's visualization data, which can be viewed with the naked eye or with the help of virtual reality equipment. Three-dimensional printing can be obtained using a new 3D printer that prints models for specific patients to better understand the pathological anatomy, or even implants or dummies for specific patients for surgical training purposes [6].

Clinical applications of these models are intended for preoperative planning, interdisciplinary communication, medical education, surgical training, and effective patient communication.

These factors are presented in the table, and also indicate the health features that need to be taken into account in such patients by cardiac surgeons.

Table 1. Concomitant diseases or conditions of patients with heart disease and risk factors that need to be considered by cardiac surgeons

Disease or condition	Risk factor
1. Age characteristics of patients	Compared to the gradually increasing age of patients with indications for transplantation in acquired heart failure, patients with congenital heart disease mostly have such indications at a younger age, with the average age of transplantation in various studies ranging from 30 to 38 years. This indicates an earlier occurrence of heart failure in patients with moderate or complex congenital heart defects. Studies show that patients with congenital heart defects accumulate a significant number of comorbidities at a young age. Careful assessment of the biological age of the patient is crucial when evaluating the indications for transplantation in patients with congenital heart disease. It should be taken into account that young patients aged 35-40 years with such a diagnosis may have a level of wear and tear of internal organs equivalent to a 70-year-old patient, both due to a pathological heart condition and due to concomitant diseases that have developed over the years.
2. Exercise tolerance	Respiratory and skeletal muscle weakness is common in young patients with complex congenital heart defects, it contributes to the development of general weakness, and may increase the predisposition to sudden, disproportionate functional decline after stressful events such as

Disease or condition	Risk factor
3. Obesity.	<p>transplants. Although patients may be young, assessment of respiratory musculature and grip strength, as well as a six-minute walk, are key components of the surgeon's assessment of the patient's health before transplantation.</p> <p>Increased body mass index (BMI) is also associated by many authors with low survival rates after transplantation. In particular, a BMI of ≥ 35 is associated with an increased risk of infection, rejection, diabetes, and cardiac allograft vasculopathy. Studies have shown that almost 15% of patients with congenital heart disease have a BMI > 30. Although reports of poorer transplant outcomes for patients with a BMI of 30-34,9 were not consistent, it is believed that a BMI of > 30 is a relative contraindication for transplantation in patients with congenital heart defects. However, it is also necessary to take into account that a number of patients with congenital heart disease are underweight and that low body weight is also associated with worse survival.</p>
4. Psychosocial problems and substance abuse.	<p>Non-compliance with the treatment regimen, active smoking or drug abuse, as well as mental health and social circumstances that may prevent patients from following the treatment regimen after transplantation, are always carefully evaluated. Although patients with congenital heart defects smoke less than the healthy population as a whole, however, more than 15% of patients with congenital heart defects have tobacco and even drug addiction. All this should also be taken into account when making a decision about heart transplantation for such patients.</p>
5. Pulmonary hypertension.	<p>Pulmonary hypertension is quite common in congenital heart defects. Patients with severe precapillary pulmonary hypertension (for example, patients with Eisenmenger syndrome) should be referred for transplantation if indicated. On the other hand, it is important to distinguish precapillary pulmonary hypertension from postcapillary or combined pre - and postcapillary pulmonary hypertension. Pulmonary hypertension often occurs in patients with a systemic right ventricle (major artery transposition (TGA) after an atrial switching procedure or a congenital corrected TGA).</p> <p>Most of these patients will have postcapillary or combined pre - and postcapillary pulmonary hypertension, compared to 3-5% with isolated precapillary pulmonary hypertension. Because pulmonary hypertension often develops with minor symptoms and is potentially reversible, the researchers recommend a low threshold for cardiac catheterization in elderly patients with systemic right ventricle, especially with elevated levels of natriuretic peptides.</p> <p>In patients with multiple sources of pulmonary blood flow, the assessment of the pulmonary vasculature can be quite complex, and decisions should be made after multidisciplinary discussions on an individual basis.</p>
6. Anatomical problems and other operations in the anamnesis.	<p>Patients with congenital heart defects have more than twice as many cases of previous sternotomy (51% vs. 24%) [5]. Although this is not a contraindication for heart transplantation as such, extensive adhesions due to</p>

Disease or condition	Risk factor
	<p>previous interventions, as well as the presence of shunt physiology, can lead to an increase in perioperative bleeding and an increase in the time of ischemia (3.8 h vs. 2.9 h) [5]. Thus, congenital heart disease is considered a risk factor for 30-day and 1-year mortality.</p> <p>Detailed axial imaging (computed tomography, triggered by an ECG) should inform the specialist about how the heart (by itself) relates to the mediastinum and the chest wall. In the same way, complex visualization of blood vessels is mandatory [7]. Detailed anatomical knowledge provides the cardiac surgeon with sufficient information to anticipate any problems that may occur during transplantation, may cause peripheral cannulation and bypass surgery, surgical planning of anastomoses, including the need to obtain large areas of systemic vessels with the transplanted organ, to identify problems with vascular access due to previous venous and arterial cannulation. Identifying and twisting large aortopulmonary collaterals can help reduce the risk of peroperational bleeding.</p>
<p>7. Liver diseases.</p>	<p>Liver diseases, including cirrhosis, are more common in congenital heart defects due to prolonged systemic venous congestion. Moreover, liver cirrhosis is associated with post-transplant mortality [1]. All patients with suspected liver dysfunction in right-sided heart disease undergo ultrasound, liver MRI (in the absence of contraindications) , and transjugular liver biopsy before being included in the transplant list, as non-invasive assessment using biomarkers is often inadequate.</p> <p>Bridged fibrosis, small liver volume, or signs of portal hypertension are indicators that the liver is unlikely to be able to cope with the requirements of a heart transplant.</p>
<p>8. Impaired kidney function</p>	<p>Kidney dysfunction is significantly more common in patients with congenital heart disease and occurs in 8% of patients with non-cyanotic congenital heart disease and 16% of patients with cyanotic congenital heart disease [6]. The pathophysiology will be somewhat similar to acquired heart failure, with chronic renal hypoperfusion and neurohormonal activation secondary to renal vasoconstriction as the main causes. An additional role can be played by cyanosis as a result of chronic hypoxia and secondary erythrocytosis with increased blood viscosity [6]. Although it can be assumed that restoring adequate blood flow after transplantation will improve kidney function, the possibility of worsening kidney function secondary to underlying conditions such as diabetes, hypertension, or primary kidney disease cannot be ruled out.</p>
<p>9. Restrictive lung diseases.</p>	<p>Chest deformities, spinal deformities, and cardiomegaly can affect ventilation and complicate the management of postoperative intensive care. Patients with congenital heart disease have a reduced tolerance to physical activity [7] and a reduced formed vital capacity of the lungs and the volume of formed exhalation [3]. It is important to take into account the number of sternotomies or thoracotomies that the patient has undergone during his life, as well as the presence of scoliosis, since this is associated with limited lung function in patients with ACHD [1].</p>

Accordingly, taking into account the above-mentioned criteria for assessing the health status of patients with congenital heart disease who are indicated for transplantation will allow us to achieve favorable results and improve the quality of life of such patients.

CONCLUSION

Thus, modern cardiac surgery technologies, rooted in the distant 50s of the twentieth century, allow us to conclude that at the present stage, surgical intervention for congenital heart defects has advanced far. Despite the challenges in this area, cardiac surgeons continue to make progress in reducing the mortality and multiple comorbidities of patients with congenital heart defects.

CONSENT

As per international standard or university standard, patients' written consent has been collected and preserved by the author.

ETHICAL APPROVAL

As per international standard or university standard written ethical approval has been collected and preserved by the author.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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