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Surgical Management for Advanced Heart Failure in Adults with Congenital Heart Disease

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Abstract

Adults with congenital heart disease (ACHD) have emerged as a new patient population that poses a variety of treatment and management obstacles. This chapter discusses the diagnosis of heart failure and treatment challenges faced by ACHD specifically addressing when to initiate mechanical circulatory support versus heart transplantation. It is evident that the ACHD population presents with a variety of unique challenges and considerations that still need to be explored. Addressing each of these issues will vastly change and improve how ACHD patients are approached from a treatment standpoint and ultimately provide more advantageous clinical options that can successfully handle the complexities presented by this population.

Keywords: transplant, VADs, heart failure, congenital heart disease

1. Introduction

Over the past several decades, adults with congenital heart disease (ACHD) have emerged as a new type of patient population that poses a variety of treatment and management obstacles. In North America alone, the prevalence of ACHD is estimated to be greater than 1 million patients [1]. The emergence of this group can be attributed to the clinical advancements that have been made in addressing these congenital disorders as they present during childhood and has enabled over 85% of children diagnosed with CHD to survive to adulthood [2]. As these patients progress into adulthood, they continue to experience complications and medical complexities associated with their CHD. The dominant complication that the ACHD

population faces is the development of heart failure, and this is currently recognized as the leading cause of death for ACHD patients [3].

In current medical practice, the gold standard for treating end stage heart failure is heart transplantation. This however remains a treatment option that is limited by donor supply. For the past several decades, the number of heart transplants that have been performed annually in the United States is between 2000 and 2500 [4]. This supply does not meet the current demands of the growing heart failure population. As the prevalence of heart failure in the ACHD population grows, the demand for heart failure treatment will continue to increase, placing further strain on the already overburdened transplant system.

In addition to the concerns associated with donor supply, ACHD patients face further burdens when seeking heart transplantation as a treatment option due to their medical complexities that are not currently accounted for in the guidelines established by UNOS. These include younger age, anatomical complexities, and decreased likelihood of an implanted mechanical assist device in comparison to the non-ACHD candidates. This, in turn, leads to a lower urgency status, longer waitlist times, and a higher incidence of ACHD patients experiencing delisting due to clinical deterioration [5].

It is evident that further evaluation of the growing ACHD population is necessary in order to provide effective management plans for the treatment of heart failure that will account for their complex circumstances. This chapter discusses current medical management, associated treatment outcomes, and future directions in the management of ACHD patients.

2. Diagnosis

The diagnosis of heart failure in ACHD is often difficult because this population may present with atypical signs and symptoms; however, diagnosis is facilitated by regular follow-up including history and physical exam, laboratory and imaging studies, and functional testing that is part of the management of these patients. Once a hemodynamic lesion is identified on imaging, correction of the lesion is usually required. If no hemodynamic lesion is present, patients are classified into two groups based on whether or not there is impaired ventricular function. Medical management of heart failure is indicated when there is impaired ventricular function without a significant hemodynamic lesion or for patients with normal ventricular function who are clinically symptomatic with either an elevated BNP or evidence of impairment of cardiopulmonary exercise testing. Regular follow-up is indicated if BNP or exercise testing is normal or for clinically, asymptomatic patients with normal ventricular function [1].

3. Treatment

3.1. Medical management

Once heart failure is recognized, medical treatment consists of a cocktail of medications including diuretics, beta blockers, renin-angiotensin-aldosterone system blockers,

mineralocorticoid receptor antagonists, digoxin, pulmonary vasodilators, calcium channel blockers, and afterload reducing agents, similar to adult-onset heart failure [1]. Treatment is tailored based on specific physiology and is outside the scope of this chapter. Other interventions include implantation of a cardioverter defibrillator [6] and cardiac resynchronization therapy [7].

3.2. Surgical management

Structural intervention is often required in patients with adult CHD and ranges from catheter based therapy to heart transplantation depending on the etiology of CHD and presentation of symptoms in adulthood. The decision to undertake surgical correction must be weighed carefully against medical management as survival decreases with an increase in the number of sternotomies [8]. Additionally, the use of blood products may cause HLA sensitization, impacting the potential for later heart transplant [9]. Cardiac surgery includes pulmonary valve/conduit replacement, closure of atrial septal defects, aortic procedures, repair/revision of tetralogy of Fallot, conversion to or revision of Fontan repair, and other valvular repair/replacements [10].

Mechanical circulatory support (MCS) assistance may be indicated for patients who develop acute heart failure resistant to maximal medical management. Extracorporeal membrane oxygenation is considered for patients who develop cardiogenic shock and often serves as a "bridge to decision" therapy in this patient population [11]. Unlike standard heart failure, ECMO is particularly useful for CHD patients who develop right ventricular failure [12]. The use of ECMO should be limited to patients who have not developed multi-organ failure as prognosis is poor in this population.

The number of chronic ventricular assist device implantations continues to increase although concentrated to relatively few centers [13]. Few patients with single ventricle morphology are implanted as most patients are classified as systemic morphological left or right ventricle [13]. Similar to ECMO therapy, long-term MCS is used as a bridge to transplant or candidacy and seldom used as destination therapy [13]. Most patients are implanted with a left VAD, but there is a higher proportion of patients compared to the acquired heart failure population who require biventricular support with either biventricular VADs or a total artificial heart [13]. Across all morphologies, axial, continuous flow pumps are more commonly used; however, there is a larger proportion of pulsatile pumps used in the ACHD population compared with those with acquired heart failure [13].

Heart transplantation is considered when estimated 1-year survival is less than 80%. The decision to list for heart transplant is complex, more so than patients with acquired heart failure, and factors influencing this decision include anatomical considerations, presence of non-heart end-stage organ failure, progressive cyanosis, degree of pulmonary hypertension, and cardiopulmonary exercise testing [14]. Patients with single ventricle morphology present particular anatomical and vascular challenges, as they often require additional surgical procedures at the time of transplant including pulmonary artery and abnormal systemic venous return reconstruction. Overall, patient selection is crucial for the success of heart transplant in adults with congenital heart disease.

4. Current outcomes for adults with congenital heart disease

4.1. Mechanical circulatory support

In the treatment of heart failure, the emergence of mechanical circulatory assist devices has become a widely accepted option for individuals who either do not meet the transplant criteria or as a bridge to transplantation. Despite their widespread use in non-ACHD patients, mechanical circulatory assist devices are not as easily applied to the ACHD population because many of these patients present with anatomical challenges such as single ventricles, vascular reconstruction of major arteries, and systemic right ventricles [2]. The complexity of anatomical variants in addition to the presence of comorbidities contributes to a higher peri-operative complication rate compared with the non-ACHD population. These adverse events include higher rates of hepatic dysfunction, respiratory failure, renal dysfunction requiring dialysis, and sustained cardiac arrhythmias [13]. When compared with a matched non-ACHD cohort, Cedars et al. found that early survival in the first 5 months post-implantation was worse in the ACHD population but comparable thereafter, and functional status and quality of life parameters were similar in both groups. They attributed these findings to the operative and perioperative factors unique to the ACHD population, particularly anatomic issues and increased likelihood of having previous sternotomies. Overall, results suggest that MCS is a good option for ACHD patients with advanced heart failure despite increased peri-operative complications and mortality as a bridge to transplant and may be a viable option as destination therapy in the future. Outcomes after MCS implantation are shown in **Table 1**.

4.2. Transplantation

ACHD patients experience a variety of disadvantages when seeking transplantation as a treatment for their heart failure. Issues such as anatomical concerns and immune status can impact their ability for transplant candidacy significantly. If these factors do not influence their ability to be placed on the transplant registry, the ACHD population experiences a higher waitlist mortality than non-ACHD patients. This can be attributed to factors such as ACHD patients typically being of a younger age and less likely to utilize mechanical circulatory assist devices due to clinical barriers. As a result, they may experience longer wait list periods, a greater incidence of death while waiting for a transplant, or delisting [5].

The outcomes for ACHD patients that are successfully transplanted vary depending on short-term versus long-term comparisons and are shown in **Table 2**. Short-term outcomes for ACHD patients, similar to outcomes after MCS, are worse than when compared to non-ACHD patients: 20–30% mortality at 30 days mortality [1]. This increased mortality rate can potentially be explained by unique challenges associated with the ACHD population such as anatomical concerns and longer times of ischemia during surgery due to the need for reconstruction during the transplant [15]. One study by Paniagua Martn et al. [16] suggests that the cause for this difference can be attributed to a higher incidence of primary graft failure in ACHD patients. Despite increased peri-operative mortality, the long-term survival for ACHD patients is outstanding, with a median survival of greater than 20 years [2].

Source	Study description	Purpose	Results
VanderPluym et al. [13]	Data entered into the Interagency Registry for Mechanically Assisted Circulatory Support (INTERMACS) from June 2006 to December 2015 was utilized. The 126 ACHD patients were categorized as follows: 63 systemic morphologic left ventricle, 45 systemic morphologic right ventricle, and 17 single ventricle.	To compare mortality between ACHD and non ACHD patients after device implantation.	The survival rate was similar between ACHD and non-ACHD patients with LVAD's.
Maly et al. [17]	Five adult patients with systemic right ventricular failure after a Mustard operation were implanted with a HeartMate II VAD.	To collect data on utilizing LVAD's as bridge to transplantation devices in patients with previously palliated transposition of great arteries.	Heart failure symptoms improved in all patients; therefore, a VAD may be a suitable treatment option in bridge to transplant for patients who are severely ill.
Everitt et al. [18]	An analysis of 9722 adults, 314 of which were diagnoses with ACHD was conducted to identify key differences in listing status and outcomes.	To analyze waitlist outcomes for ACHD versus non ACHD patients in heart transplantation.	Adults with CHD were much less likely to have a VAD (5 versus 14%) and were more likely to be given a lower urgency status. These patients were also more likely to experience cardiovascular related death with waiting to undergo heart transplantation (60 versus 40%). The utilization of VAD's should be explored to determine if survival for ACHD patients can be improved.
Shah et al. [19]	A retrospective analysis of six ACHD patients who underwent VAD implantation.	To provide data for ACHD patients with VAD implantation.	Five patients survived to discharge: one patient was successfully transplanted, one patient survived 262 days; one patient received 988 days of therapy while awaiting transplantation as of December 1, 2012; and two patients who received VADs as destination therapy received 577 and 493 days and were still alive as of December 1, 2012. VAD implantation is a viable option for therapy in ACHD patients in either bridge to transplant or bridge to destination therapy.
Newcomb et al. [20]	An ACHD patient with failing Fontan circulation was implanted with an LVAD device and went on to have a successful heart transplantation 5 months later.	A case study that discusses the outcome of an LVAD implantation in an ACHD patient with a failing Fontan circulation as bridge to transplant therapy.	This case report suggests that LVAD's can become useful in patients with ACHD, particularly those with failing Fontan circulation as either bridge to transplant or bridge to destination therapy.

Source	Study description	Purpose	Results
Morris et al. [21]	A presentation of a case in which LVAD implantation was utilized as therapy option in an ACHD patient that presented with failure of the systemic ventricle.	A case study of LVAD implantation in an ACHD patient.	Utilization of the LVAD therapy significantly improved the patients cyanosis and ventricular function. This suggests that patients with ACHD could benefit from utilizing LVAD therapy.
Stewart et al. [22]	Two ACHD patients were successfully bridged to transplantation utilizing LVAD therapy. Their deterioration leading to the need for transplant can be attributed to their deteriorating right ventricular failure.	To explain the case reports of two ACHD patients who received LVAD bridge to transplant therapy.	This report suggests that LVAD therapy is a viable option for bridge to transplantation in ACHD patients that present with right ventricular failure.
Gelow et al. [23]	A retrospective study of 1250 ACHD patients reported in the UNOS database from 1985 to 2010 in which these patients were compared to non –ACHD patients in terms of VAD use at listing, listing status, status upgrades and reasons for upgrades.	To determine the relationship that exists between VAD implantation and successful transplantation in patients that are listed for heart transplant.	It was noted that the use of VAD's in ACHD patient was less at both the time of listing and transplantation. This decreased usage of VAD therapy in ACHD patients contributes to lower listing status and organ allocation.
Joyce et al. [24]	Three adult patients that had congenitally corrected transposition of the great arteries underwent LVAD implantation as a therapy option for their end stage heart failure.	A case report of three ACHD patients that underwent LVAD implantation.	LVAD implantation can be successfully completed in ACHD patients when placed under echocardiographic guidance. This offers an additional therapy option for ACHD patients.
Maxwell et al. [25]	Data collected from September 1987 to September 2012 by the Scientific Registry of Transplant Recipients was utilized to compare the following between MCS and non-MCS ACHD patient populations: procedural, outcome and survival.	To analyze the pretransplant effects of mechanical circulatory support on posttransplant outcomes in the ACHD population.	In the ACHD patient population, those with MCS are associated with higher transfusion rates and length of stay however, they do not have less favorable outcomes post-transplant when compared to non-MCS ACHD patients.

Table 1. Outcomes after mechanical circulatory support device implantation in adults with congenital heart disease.

Source	Sample/study description	Purpose	Results
Irving et al. [26]	Outcomes were reviewed from 38 cardiac transplants performed in 37 patients from 1988 to 2009 using medical records and transplant databases. 41% had univentricular and 59% had biventricular physiology.	To explore data on outcomes of cardiac transplantation in the ACHD patient group.	Operative mortality for ACHD patients following cardiac transplantation is higher than for other diagnostic groups. However, long term survival is noted to be good and comparable to non ACHD patients.

Source	Sample/study description	Purpose	Results
Patel et al. [27]	Data reported to UNOS from 1987 to 2006 was reviewed and categorized to compare adults with CHD versus other diagnoses in heart transplantations. 2% of the individuals in this study period had CHD.	To evaluate the post transplantation prognosis in adults with CHD.	The 30-day mortality rate is elevated in the ACHD population: 16 versus 6%. However, there is not a statistical significance in the 5 and 10-year survival rates for ACHD patients in comparison to non-ACHD patients.
Taylor et al. [28]	Data from heart transplantations performed from 2001 to 2003 was utilized to calculate survival rates by the Kaplan–Meier method. Adults with CHD represented 2.7% of the cohort.	To evaluate the survival outcomes for patients post heart transplantation.	Having a diagnosis of ACHD is one of the most powerful predictors of 1-year mortality. But at 10 years it is associated with a marked survival advantage conditional on a 3-year survival independent of age.
Lamour et al. [29]	The post-transplantation outcomes for 24 adults with CHD were analyzed utilizing the Kaplan–Meier statistical method to estimate survival functions for patients with CHD versus all others and patients with CHD versus matched controls.	To analyze the survival rate of adult patients with CHD post cardiac transplantation in comparison in those without CHD.	The survival rate for patients with ACHD post-transplantation was 79% at 1 year and 60% at 5 years. A difference between this population and the control populations was not present.
Davies et al. [8]	A retrospective study of patients listed for primary transplantation between 1995 and 2009 was conducted. 2.5% of these patients were adults with CHD.	To evaluate the survival of adults with CHD after listing and transplantation.	The early mortality rate (30 day) among ACHD patients was high (reoperation 18.9 versus 9.6%; nonreoperation 16.6 versus 6.3%), but at 10 years the survival rate was equivalent with non-ACHD patients (53.8 versus 53.6%)
Bhama et al. [30]	A retrospective analysis was conducted from January 2001 to February 2011. 19 patients with ACHD were compared to 428 patients with non-ACHD who underwent transplantation.	To evaluate the survival outcomes of cardiac transplantation in adults with CHD in a contemporary cohort.	There was no significant difference in survival of ACHD versus non-ACHD at 30 days (89 versus 92%), 1 year (84 versus 86%), or 5 years (70 versus 72%).
Karamlou et al. [31]	A review of heart transplantation patients from 1990 to 2008 reported to UNOS was conducted. A total of 8496 patients were evaluated, of which 575 had ACHD.	To investigate outcomes and risk factors for mortality and retransplantation for the ACHD population in comparison to the non-ACHD population.	The overall post-transplantation mortality and retransplantation rates were significantly higher for patients with ACHD mainly due to an early hazard phase.
Burchill et al. [32]	A retrospective study was conducted on patients who were identified in the registry of ISHLT between 1985 and 2010. The Kaplan–Meier method was used to conduct a survival comparison. 2.2% of patients transplanted in this cohort had a diagnosis of ACHD.	To examine survival, causes of death and predictors of early (<1 year), mid-term (1–5 years) and later (0.5 years) mortality in ACHD patients who received cardiac transplants.	Early mortality rates for the ACHD population was high in comparison to the non-ACHD transplant recipients (10 versus 4%). The long-term survival rates for ACHD patients who survived the early hazard phase was superior to the non-ACHD patients.

Source	Sample/study description	Purpose	Results
Paniagua Martin et al. [16]	Survival outcomes in a total of 3166 patients were included: 1888 IHD, 1223 IDCM, and 55 ACHD.	To analyze the survival probability between different subgroups with ACHD.	The early mortality rating associated with ACHD can primarily be attributed to the presence of primary graft failure. The frequency of primary graft failure in ACHD was 23%, versus 17% in IHD and 13% in IDCM. The following is the frequency of early mortality rates: 25% CHD, 14% IDCM, 16% IHD.
Singh et al. [33]	Adults who underwent heart transplantation in the United States between January 2007 and June 2009 were utilized to determine and validate the risk prediction model. This efficiency of this model was further assessed by evaluating the performance in patients from July 2009 to October 2010 receiving heart transplants.	To develop a risk prediction model for posttransplant in hospital mortality in heart transplant patients.	The model determined that the ACHD diagnosis is correlated with an odds ratio of 4.18 for early in hospital mortality post heart transplantation.
Karamlou et al. [34]	A comparison among in hospital deaths between ACHD patients that possessed either 1 V or 2 V anatomy was conducted retrospectively from 1993 to 2007 through data gathered in the Nationwide Inpatient Sample (NIS).	To determine if there is an associated with early death post heart transplantation in patients who possess 1 V anatomy in ACHD.	ACHD patients that possess 1 V anatomy are associated with a higher death incidence post heart transplantation. Transplantation registries should include specific ACHD diagnoses due to the evident difference in associated outcomes.

Table 2. Outcomes after heart transplantation in adults with congenital heart disease.

Regardless of this data, outcomes for patients with ACHD after transplantation vary depending on their initial diagnosis. As there are a variety of clinical manifestations of ACHD, assessing prognostic values remains challenging and therefore individuals should be evaluated thoroughly prior to transplant consideration.

5. Conclusion

Further investigation into the ACHD population is essential in order to effectively manage their unique medical concerns as this patient group continues to expand. This investigation must occur from multiple points in order to ensure the variety of distinct challenges presented by this population are adequately addressed. Specifically, there are four areas this chapter suggests future research efforts should focus on in order to provide the most advantageous information for medical management:

- The cause of increased early mortality rates in heart transplant operations for ACHD patients. After thorough review of the current literature, it is evident that ACHD patients experience

higher early mortality rate post heart transplantation in comparison to non-ACHD patients. However, at this point in time little research has been focused on identifying the clinical source for this mortality contrast. It is essential that research efforts focus on seeking out the root of this disparity in order to work towards minimizing the presence of this current complex outcome. Doing so will supply the medical community with more accurate predictors of mortality when seeking heart transplantation as treatment for these patients and provide better outcomes to those who undergo this type of medical management.

• Determining the appropriate timing/type of interventions to utilize for this clinically diverse group. Due to the clinical diversity that exists within the ACHD patient populations applying standardized treatment regimens remains challenging. Case studies exploring how to effectively manage different anatomical morphologies currently exist but this aspect of research still remains relatively unexamined and information specifically regarding timing is rather limited. Increasing the knowledge in terms of how to effectively approach treatment in ACHD patients in terms of when and how to intervene will assist in decreasing the complexity of approaching a therapy regimen and provide stronger evidence to provide the best possible clinical outcomes for these patients.

• Re-evaluating how ACHD patients are listed into the transplant registries. With the current listing guidelines ACHD patients are at a significant disadvantage in terms of their likelihood of being successfully transplanted. As of now, ACHD patients are more likely to experience a lower listing status with their initial listing than non-ACHD patients. In addition, ACHD patients experience a high rate of delisting after 1 year due to a decline in their worsening condition. These patients are placed at an even further disadvantage because they may not be candidates for mechanical circulatory support due to anatomical constraints. Therefore, they are unable to utilize the placement of these devices to prolong their survival to successfully reach transplantation, or utilize the benefits of attaining a higher listing status associated with these interventional therapies. The current listing criteria for heart transplantation is a cause of serious concern when considering ethical and effective medical management for patients with ACHD. There is an urgent need for re-evaluation of these current guidelines to occur in order to take into consideration the unique medical challenges presented by this growing population that will continue to rely on heart transplantation as one of their main treatment possibilities in the future.

• Exploring the use of MCS as destination therapy in addition to bridge to transplantation. The utilization of these devices for treatment in ACHD patients has previously focused on their usage as bridge to transplant therapy. However, with the increasing demand for heart transplantation, it is imperative that other therapy options are considered for ACHD patients. More recently, the use of MCS has been considered as destination therapy for this group of patients. Current research indicates that there is potential for pursuing this line of treatment option for a variety of ACHD subgroups. Doing so would provide an effective treatment option for these patients and relieve some of the current burden on the transplant system.

It is evident that the ACHD population presents with a variety of unique challenges and considerations that still need to be explored. Addressing each of these areas mentioned above

will vastly change and improve how ACHD patients are approached from a treatment stand-point and ultimately provide more advantageous clinical options that can successfully handle the complexities presented by this population.

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