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Organ Allocation In Adults With Congenital Heart Disease Listed For Heart Transplant: The Impact Of Ventricular Assist Devices

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Abstract

Background—Adults with congenital heart disease (CHD) listed for heart transplantation infrequently are supported with ventricular assist devices (VADs). This may disadvantage their priority for organ allocation. We sought to determine the relationship between VAD implantation and successful transplantation among patients listed for heart transplant.

Methods—Adults with CHD patients (N=1,250) were identified in the United Network for Organ Sharing (UNOS) database from 1985 – 2010 and compared to patients without congenital etiology for heart failure (N=59,606). VAD use at listing, listing status, status upgrades, and reasons for upgrade prior to transplant were trended at 5 year intervals and appropriate statistical comparisons were made between groups.

Results—Since 1985, VAD use prior to transplant has increased significantly in patients without CHD but not in CHD patients (17% vs. 3% in 2006–2010, $p<0.0001$). CHD patients were more likely to be listed as status 2, compared to those without (66% vs. 40%, $p<0.001$ for 2006–2010), and less likely to be upgraded to status 1 after listing (43% vs. 55%, $p=0.03$). Among those upgraded to status 1, CHD patients were less likely to have a VAD at transplant than those without (3% vs. 18%, $p=0.005$). VAD use was more likely to result in death in CHD patients.

Conclusions—VAD use is less common in CHD patients than patients without CHD, both at the time of listing and transplantation. Reduced VAD use appears to contribute to lower listing status and organ allocation. These differences have grown more disparate over time. Separate criteria for organ allocation for CHD patients may be justified.

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Conflict of Interest

None of the authors have any relevant financial relationships to disclose.

Background

Adult congenital heart disease (CHD) patients are a growing population because of improved survivorship through childhood. They remain vulnerable to myocardial dysfunction and clinical heart failure,¹ a major cause of death in these individuals.^{2, 3} Because of this, adult CHD patients are a growing proportion of patients referred for heart transplantation.⁴ Despite the anatomic and physiologic challenges,⁵ favorable longterm transplant outcomes have been reported.^{6, 7}

The use of ventricular assist devices (VAD) as a bridge to transplant has become more commonplace, particularly since the introduction of continuous flow pumps.^{8, 9} Extension of this practice to CHD patients, however, has been slower. Data from the United Network for Organ Sharing (UNOS) Standard Transplant and Research dataset has demonstrated that, compared to those without CHD, listed CHD patients are less likely to have a VAD or other mechanical support, and longer wait times in status 2.^{10, 11} Consequently, cardiovascular mortality on the heart transplant wait list is higher in CHD patients.

In 2006, a universal policy change was made in US organ allocation, such that status 1 patients outside the local referral area were offered organs before local status 2 patients. This change decreased the number of overall transplants for status 2 patients.^{4, 12, 13} Because VAD implantation is a qualification for status 1 listing, lower VAD implantation rates in listed patients with CHD may result in lower priority status and reduced organ allocation for these patients.¹² We sought to determine the impact of VAD utilization on listing and heart allocation for CHD heart transplant candidates by following trends over time.

Methods

Patient Population

Patient-level data were obtained from UNOS, a US registry of transplant listing, organ allocation, and outcomes maintained continuously since 1985. Our Institutional Review Board approved our use of these data, and the requirement for individual consent was waived. We excluded patients who were <18 years old at the time of listing, listed for double organ transplantation, or listed for re-transplantation. The remaining patients were classified as CHD or without CHD patients based on the stated etiology of their heart failure.

Variables obtained included age, sex, listing status, inotrope use, and VAD implant at the time of listing. Patients who were status 2 at listing but status 1 (1, 1A or 1B) at the time of organ offering were considered to have had a status upgrade. Patients in whom inotropic support was provided at time of transplant but not at listing were considered upgraded on the basis of inotrope use. Similarly, patients in whom VAD was present at transplant but not at listing were considered upgraded on the basis of VAD placement. Both were expressed as a percentage of patients upgraded. These were not mutually exclusive, nor did they account for all upgrades. All VADs were included together regardless of designation as “right” or “left,” given the potential incongruity of nomenclature for systemic vs. sub-pulmonic ventricles. Missing values for VAD fields were assumed to indicate no VAD support was present.

Data were analyzed by 5-year incremental eras starting from 1985 and were based on the listing date. Groups were compared using SPSS for Mac (Version 18) using Chi-square testing for categorical variables, and Students t-test for continuous variables. $P < 0.05$ was considered statistically significant. All authors have had access to the primary UNOS data,

have read and agree to the manuscript as written, and take full responsibility for the integrity of the reported findings.

Results

Of 78,470 individuals in the database, 40,785 were excluded (including 13,177 pediatric patients, 4,068 listed for multiple organs simultaneously, and 2,389 listed for redo transplant, not mutually exclusive), leaving a study population of 60,856. From these, we identified 1,250 CHD patients (36% female), and 59,606 without CHD (22% female). CHD patients in general were younger at listing (33.5 ± 12.5 years vs. 51.4 ± 11.2 years, $p < 0.001$), as expected from previous studies.^{10, 11} Peak VO_2 was not significantly different (12.6 ± 3.2 vs. 11.6 ± 3.5 ml/kg/min). The majority of CHD patients were classified as “CHD with unknown surgery” ($N=828$), with 372 identified as “CHD with surgery” and 47 as “CHD without surgery.”

Adults listed for transplantation for both groups is given by era (table 1). The percentage of listed patients who are transplanted has declined over time, with a larger drop in CHD patients to 50% (95% CI 45 – 56%) vs. 62% (95% CI 61 – 63%) of patients without CHD for the most recent era ($p < 0.001$).

The percentage of patients initially listed as status 2 has not changed in the CHD group for the past three eras (Figure 1). However, this percentage has gradually dropped for those without CHD to 40% in the most recent era (95% CI 39 – 41%, $p < 0.001$ vs. CHD). The number of patients transplanted after initial listing in status 1 has not changed significantly over time for either group. Yet there has been a decline in transplantation from status 2 since 2006 for both groups (Figure 2). Currently the proportion of patients transplanted from status 2 is 33% (95% CI 24 – 42%) for CHD, and 41% (95% CI 39 – 44%) for those without CHD ($p < 0.0001$ for both compared to prior era).

For patients without CHD, VAD use rose steadily to 17% (95% CI 16–17%) at listing and to 17% (95% CI 16.5 – 17.8%) at transplant over the study period ($p < 0.001$ for change from 1985–1990 for both listing and transplant, Figure 3). Strikingly, there has not been a significant change in VAD utilization in CHD patients over this same time period.

The frequency of status upgrades while listed is shown for both groups (Figure 4). A similar percentage of patients were upgraded from status 2 at listing to status 1 at transplant for both CHD vs. those without CHD during all eras except 2006 – 2010, when there was a significantly higher percentage upgraded among patients without CHD group (55%, 95% CI 53 --56%) compared to the CHD group (43%, 95% CI 36 – 51%, $p=0.003$ vs. those without). Of those upgraded, the percent of patients upgraded with interval institution of inotrope support did not differ between groups (Figure 5). This contrasts with the percent of patients upgraded because of interval VAD implantation, which was significantly higher in those without CHD for each era from 1996 (Figure 6). For 2006 – 2010, 18% of patients without CHD upgraded had a VAD at the time of transplant (95% CI 16 -- 20%) vs. only 3% in CHD patients (95% CI 0-- 6%, $p=0.005$).

The outcome of VAD patients at listing for the entire study period is shown (Table 2). For non-CHD, 68% (95% CI 67 – 70%) were transplanted, and 26% (95% CI 24 – 27%) died or were unstable. The remaining patients (6%) were removed for other reasons including transfer out of area, refusal, clinical improvement, or other. By comparison, for CHD patients, 48% (95% CI 33 – 63%) were eventually transplanted, 41% (95% CI 26 – 55%) had died or were too unstable for transplant, and 12% were removed from listing ($p=0.015$ for CHD vs. without CHD).

Discussion

Our analysis of the UNOS data demonstrates that CHD patients are: 1) less likely to have a status upgrade while listed, 2) less likely to receive a VAD after being listed, 3) less likely to receive an allograft while listed, and 4) more likely to die or become too unstable rather than be transplanted if a VAD is present. Additionally, since 2006, there has been a considerable decline in transplantation from status 2 among patients without CHD and especially with CHD, reflecting the allocation policy shift.

The observed low utilization of VAD among CHD patients can be interpreted in several ways. One possibility is that less severe heart failure is present in CHD patients listed for transplant. This seems unlikely since CHD patients had similar exercise capacity and worse cardiovascular outcomes while listed.¹¹ A second more likely explanation is that increased anatomic complexity limits implantation. Based on single-center publications, the majority of congenital heart transplants are done in patients with transposition of the great arteries with a systemic right ventricle,^{14–16} whereas most VADs are designed for implantation in a morphologic left ventricle. An alternative explanation is that CHD patients suffer severe circulatory failure, but not from ventricular systolic dysfunction. The patients most likely to fit this category are single-ventricle patients palliated with a Fontan procedure, who comprise a considerable portion of transplanted CHD individuals.^{14–16} Many single ventricle patients develop significant morbidity from chronically elevated systemic venous pressure, including protein-losing enteropathy (PLE), malnutrition, ascites, coagulopathy, liver cirrhosis and desaturation through veno-venous collaterals. All of these occur in the context of normal systolic function of the systemic single ventricle. VAD use in the systemic circulation is therefore not a suitable solution for “Fontan failure.”^{17, 18} To support the pulmonary circulation, VAD implant requires complex cannulation,¹⁹ and is currently reported only anecdotally,^{20, 21} often without successful outcome.²² Novel pump designs have been proposed specifically for Fontan patients to address this problem,^{19, 23} with the expectation that successful utilization may favorably impact transplant candidacy and outcome.

Status upgrades in both groups were also attributable to inotropic support. However, dobutamine does not increase stroke volume in Fontan or systemic RV patients.^{24–26, 27, 28} Thus inotropic support as a criterion for heart failure severity and heart allocation priority may also not be applicable to this population.

Given the scarcity of organs, should transplant priority be given to patients with lower risk of perioperative complications? CHD patients represent a dilemma; higher risk may be balanced by high potential benefits. While early mortality in specific subsets is higher,²⁹ long-term survival is excellent and comparable to outcome in recipients without CHD. Despite these realistic concerns, in a contemporary cohort from a single center, there was no difference in post heart transplant survival at one month, one year or five years between patients with CHD or without CHD.³⁰ As such, transplantation remains an important therapy for CHD patients. They are generally younger than patients without CHD and their transplant survival benefit may add years to their adulthood.^{6, 7} PLE, for example, contributes substantially to perioperative mortality,³¹ yet transplantation is curative.^{18, 32, 33} Despite the favorable long-term outcomes, one could argue that VAD use stabilizes the patients prior to transplant, and that patients with higher perioperative risk, such as single ventricle patients,²⁹ should not necessarily have priority. Yet CHD patients in whom a VAD is not an option may improve their perioperative outcomes by earlier transplantation prior to progression of comorbidities and concomitant risk.

Clinical Relevance

The 2006 allocation policy change created a disadvantage for status 2 patients, who now wait longer for an organ. Because VAD implant defines a status 1A or 1B patient, there is a “cutting in line” effect for those with VADs who have a head start in allocation for this precious resource. VAD use offers recipients a period of potential stability making them better suited for eventual transplant.³⁴ Non-VAD candidates do not have this safety net. In light of the disparity, we agree with others that for certain sub-populations such as those with CHD, inapplicable or less-effective therapies such as VAD implantation or inotrope dependence should not be so heavily weighted in the determination of organ allocation.^{10–12} It has been recently shown that the current status definitions that favor VAD patients are disproportionate to their actual mortality risk relative to those without VAD.³⁴ Collectively the data support a reappraisal of allocation procedures to render the organ allocation process more equitable. Allocation priorities should balance the risks of mortality without transplant as well as perioperative risks with the relative long-term benefits of transplantation in younger patients.

Limitations

Inherent limitations of this study include the retrospective design from a dataset not necessarily tailored for CHD patients. There is no method to validate the reported CHD diagnosis within the registry. Our methods required assumption that missing data, particularly for VAD use, implied that therapy was not given. While we cannot formally validate this assumption, we know from the Interagency Registry for Mechanically Assisted Circulatory Support (INTERMACS) dataset on implanted devices instigated in 2005, that only 21 VADs were registered in CHD patients, which is comparable with the data we reported in this study. We did not differentiate right versus left VAD because of lack of standardized nomenclature for congenital defects and uncertainty as to which circulation was being supported. We did not study children, in whom unique issues are often raised, though bridging to transplant with VAD has been done successfully.^{21, 35}

Conclusions

CHD poses new challenges to heart failure management and transplantation. Use of mechanical support as a bridge to transplantation in the CHD population is currently limited, and highlights the need for VAD innovation and the development of surgical techniques for VAD implantation in anatomically complex CHD patients. Given the significant challenges of VAD use in this population, the feasibility of increasing VAD support in patients with CHD remains in question. Orthotopic heart transplant has been shown to be an effective therapy in these individuals. Given this, current allocation policies that favor patients without CHD may not be fair. Though the proportion of CHD patients undergoing transplant is small, national standards should account for their unique limitations and allow equitable organ allocation based on both risk and potential benefit.

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Status 2 at Listing

Figure 1.

Percentage of patients from each group listed in status 2 for both groups as a function of era. CHD = congenital heart disease. Controls are adult patients without CHD.

Transplanted from Status 2

Figure 2.

Percentage of patients from each group transplanted from status 2 as a function of era. CHD = congenital heart disease, control are those without CHD. P values are for comparison between eras for both patient groups.

VAD use by Era

Figure 3.

Percentage of patients with implanted ventricular assist devices at listing and transplant for both congenital heart patients (CHD) and those without (controls), shown by era. P values are for comparison between patient groups for VAD use at transplant.

Status Upgrade While Listed

Figure 4.

Percentage of patients from either group who were upgraded from status 2 at listing to status 1 at time of organ offering as a function of era. CHD = congenital heart disease.

Status Upgrade from Inotrope Use

Figure 5. Percentage of patients upgraded from status 2 with interval requirement of inotropic support. CHD = congenital heart disease.

Status Upgrade from VAD Placement

Figure 6. Percentage of patients upgraded from status 2 with interval ventricular assist device implant (C). CHD = congenital heart disease.

Table 1

Adults listed for heart transplantation by era.

Era	CHD Listed (N)	CHD Transplanted (%)	95%CI	Without CHD Listed (N)	Without CHD Transplanted (%)	95%CI
1985–1990	69	100	(100–100)	6,712	85	(85 – 86)
1991–1995	204	77	(71 – 82)	14,004	70	(69 – 71)
1996–2000	293	57	(52 – 63)	15,414	61	(60 – 62)
2001–2005	366	67	(62 – 72)	11,825	67	(66 – 67)
2006–2010	318	50	(45 – 56)	11,651	62 [*]	(61 – 63)

Number of adults listed for transplantation by era, together with percent transplanted with 95 confidence limits), for adults with congenital heart patients (CHD) vs. those without.

^{*} denotes p<0.001 for comparison of patients CHD vs. without.

Table 2

Outcome of all patients with VADs at the time of listing

	CHD			Without CHD		
	N	%	95% CI	N	%	95% CI
Transplanted	20	48	(33–63)	3001	68	(67–70)
Died/unstable	17	40	(26–55)	1128	26	(24–27)
Removed/other	5	12	(2–22)	255	6	(5–7)

CHD = congenital heart disease. Removed/other category includes those with clinical improvement, transfer away from the transplant center, refusal of transplant, or removed in error.