REVIEW

Heart transplantation in adults with congenital heart disease

Transplantation cardiaque chez les congénitaux adultes

Lucile Houyel a, *, Ngoc-Tram To-Dumontier b, Yannick Lepers b, Jérôme Petit a, Régine Roussin a, Mohamed Ly a, Emmanuel Lebret a, Elie Fadel c, Jürgen Hörer a, Sébastien Hascoët a

a Service de Chirurgie des Cardiopathies Congénitales, Hôpital Marie-Lannelongue-M3C, Le Plessis-Robinson, France
b Service de Cardiologie, Hôpital Marie-Lannelongue, Le Plessis-Robinson, France
c Service de Chirurgie Thoracique, Hôpital Marie-Lannelongue, Le Plessis-Robinson, France

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Summary With the advances in congenital cardiac surgery and postoperative care, an increasing number of children with complex congenital heart disease now reach adulthood. There are already more adults than children living with a congenital heart defect, including patients with complex congenital heart defects. Among these adults with congenital heart disease, a significant number will develop ventricular dysfunction over time. Heart failure accounts for 26–42% of deaths in adults with congenital heart defects. Heart transplantation, or heart–lung transplantation in Eisenmenger syndrome, then becomes the ultimate therapeutic possibility for these patients. This population is deemed to be at high risk of mortality after heart transplantation, although their long-term survival is similar to that of patients transplanted for other reasons. Indeed, heart transplantation in adults with congenital heart disease is often challenging, because of several potential problems: complex cardiac and vascular anatomy, multiple previous palliative and corrective surgeries, and effects on other organs (kidney, liver, lungs) of long-standing cardiac dysfunction or cyanosis, with frequent elevation of pulmonary vascular resistance.

Abbreviations: ACHD, adult congenital heart disease; BTT, Blalock-Taussig-Thomas; CHD, congenital heart disease; NT-proBNP, N-terminal pro-B-type natriuretic peptide; PA, pulmonary artery; PLE, protein-losing enteropathy; PVR, pulmonary vascular resistance; TGA, transposition of the great arteries; VO2max, maximal oxygen uptake.

* Corresponding author at: Service de Chirurgie des Cardiopathies Congénitales, 133 avenue de la Résistance, 92350 Le Plessis-Robinson, France.

E-mail address: l.houyel@ccml.fr (L. Houyel).

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Resistance. In this review, we focus on the specific problems relating to heart and heart–lung transplantation in this population, revisit the indications/contraindications, and update the long-term outcomes.

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Background

An increasing number of children with complex congenital heart disease (CHD) now reach adulthood due to improvements in cardiac surgery and postoperative care. Today, there are more adults than children with a congenital heart defect, including complex CHD [1]. A significant number of these adults will develop ventricular dysfunction. Heart failure accounts for 26–42% of deaths in adult congenital heart disease (ACHD) [2,3]. Heart transplantation, or heart–lung transplantation in Eisenmenger syndrome, then becomes the ultimate therapeutic possibility for these patients. This population is at high risk of death after heart transplantation, although their long-term survival is similar to that of patients transplanted for other reasons [4,5]. Heart transplantation in ACHD remains challenging because of the complex cardiac and vascular anatomy, multiple previous palliative and corrective surgeries, and effects on other organs (kidney, liver, lungs) of long-standing cardiac dysfunction or cyanosis, with frequent elevation of pulmonary vascular resistance (PVR).

Heart transplantation

Indications and population concerned

Every patient with a CHD that is repaired or palliated in infancy or childhood can develop late myocardial dysfunction, even those with CHDs that are considered “minor”, such as atrial or ventricular septal defects. However, two main categories of patients are particularly exposed to late cardiac dysfunction: those with a systemic right ventricle (congenitally corrected transposition of the great arteries [TGA] or TGA repaired by an atrial switch technique [Mustard or Senning]) and patients with a functionally univentricular heart, non-operated or palliated by various techniques, including total cavopulmonary connection (Fontan-type circulation).

The profile of this population is evolving. Over 25 years (1988–2012), among a series of 2257 heart transplantations in three centres in Paris, including 100 performed in ACHD, the proportion of univentricular hearts did not change, but the number of patients with a failing systemic ventricle increased, because of the increasing number of TGAs with atrial switch performed in infancy [6].

The definition of cardiac failure in ACHD is relatively imprecise [7]. The definition of the Heart Failure Society of America has recently been endorsed by the American Heart Association: "heart failure is a syndrome characterized by either or both pulmonary and systemic venous congestion and/or inadequate peripheral oxygen delivery, at rest or during stress, caused by cardiac dysfunction” [8]. However, other definitions have been proposed, essentially because some patients with ACHD have a low aerobic capacity and/or elevation of N-terminal pro-B-type natriuretic peptide (NT-proBNP) at baseline, although asymptomatic [7]. Threshold values for these two variables have been
proposed: NT-proBNP ≥ 100 pg/mL and maximal oxygen uptake (VO₂max) < 25 mL/kg/min [9]. In addition, atrial arrhythmia or sudden death can be the first manifestation of cardiac failure in ACHD. Arrhythmias are intimately related to cardiac failure, and can be either the cause or the consequence of this condition [7]. Among the other causes, prolonged cyanosis, leading to inadequate tissue oxygenation, may play a major role in the occurrence of late cardiac dysfunction in ACHD [7].

The prevalence of cardiac insufficiency in ACHD reaches 20–30% after a Mustard or Senning operation, 32% in congenitally corrected TGA and 40% in univentricular hearts [7]. Heart failure is now recognized as a leading cause of mortality, morbidity and hospitalization in ACHD [2,3,10–13].

Timing of insersion on the waiting list is particularly difficult to determine in ACHD, because life expectancy is even more difficult to predict in this population than in other candidates for heart transplantation [14]. This is especially true for patients with Eisenmenger syndrome, as their survival is better than in pulmonary hypertension from other causes, and because of the complications of lung and heart–lung transplantation [14].

Pretransplantation evaluation

Pretransplantation evaluation is not basically different for patients with ACHD compared with other heart transplant candidates. This evaluation aims to confirm the indication, and to search carefully for contraindications [19]; it is performed within the transplant centre, during a short (3–4 days) hospitalization, and includes cardiac catheterization and cardiac angiography, a computed tomography scan or magnetic resonance imaging to assess the individual anatomy with regard to additional procedures during transplantation (tunneлизation of caval veins, pulmonary artery [PA] reconstruction, aortic arch procedures), arterial Doppler examinations, a battery of blood tests and an evaluation by a psychologist and/or psychiatrist. Detailed information about heart transplantation and its risks and consequences is then given to the patient, during one or several conversations between the transplant physician and the surgeon, and the patient and their family. This information should be as complete and objective as possible, to include the patient in the therapeutic project, to obtain perfect compliance with immunosuppressive therapy after transplantation.

Confirmation of the necessity for heart transplantation

Heart transplantation is indicated in the presence of symptomatic ventricular or biventricular dysfunction, which has already required one or several hospitalizations for congestive cardiac failure or low cardiac output, despite maximal medical treatment. This applies to patients in New York Heart Association stage III or IV, with a major functional impairment demonstrated by VO₂max or a 6-minute walk test. The criteria commonly required to be obtained in these functional tests by patients without CHD are VO₂max < 12 mL/kg/min and 6-minute walk test <50% of predicted value. However, VO₂max < 50% of theoretical values or >25 mL/kg/min may be more appropriate in young patients [9].

Search for contraindications

Some contraindications are absolute [15,16]: pulmonary vascular resistance >4 Wood units (better if measured by the Fick method) and/or transpulmonary gradient (the difference between mean PA pressure and mean capillary pressure) >12 mmHg [17]; cancer in evolution; uncontrolled chronic infection; severe peripheral vascular disease; serious psychiatric problems likely to result in non-compliance with immunosuppressive therapy; mental or behavioural disabilities; addiction to alcohol and other drugs; and non-compliance with treatment, whatever the reason.

Other contraindications are relative [15]: obesity (body mass index > 30 kg/m²); chronic obstructive pulmonary disease; severe chronic renal insufficiency with glomerular filtration rate < 30 mL/min (in this case, combined cardiac and renal transplantation should be considered); diabetes with severe peripheral disease; and active smoking.

Specific problems in adults with congenital heart disease

Pulmonary hypertension

Many patients with ACHD develop pulmonary vascular disease because of long-lasting elevation of systemic ventricular filling pressures [18]. Right heart catheterization is mandatory in all transplant candidates, especially in adults as part of the preoperative evaluation, and should be repeated every 6 months or every year until transplantation, depending on the degree of PA pressure [15]. Elevated PVR > 4 Wood units and transpulmonary pressure gradient > 12 mmHg are considered irreversible if they do not decrease after 24 or 48 h of specific treatment (diuretics, inotropes, inhaled nitric oxide) [15], and are thus commonly considered to be a contraindication to heart transplantation. Recently, the International Society of Heart and Lung Transplantation revised and raised these levels up to PVR > 5 Wood units and transpulmonary pressure gradient > 16–20 mmHg [19]. Beyond these values, there is a major risk of acute right ventricular dysfunction in the immediate postoperative period, resulting in an increase in postoperative mortality and morbidity. In this case, to propose heart–lung transplantation instead of cardiac transplantation alone remains a difficult decision, because of the higher postoperative risk and the lower survival rate of this surgery. However, advances in pulmonary arterial antihypertensive treatments can challenge the irreversibility of elevated PVR as defined above. When PVR is too high to allow insertion on the waiting list for heart transplantation, oral pulmonary arterial hypertension-specific drug therapy may be used in an attempt to reduce it before listing. Isolated cases of successful heart transplantation after normalization of PVR under oral sildenafil–bosentan combination therapy have been reported [20,21]. One of our patients, a 43-year-old woman with corrected TGA (S, L, L) associated with ventricular septal defect, pulmonary atresia and dextrocardia, who had two Blalock-Taussig-Thomas (BTT) aortopulmonary shunts in infancy (bilateral thoracotomy), was initially treated with bosentan because of a mean PA pressure measured at 32 mmHg at heart catheterization.

One year later, heart transplantation was considered, but the mean PA pressure was 45 mmHg at catheterization, and the patient was referred for heart–lung transplantation. However, heart–lung transplantation was discarded because of previous thoracotomies. Sildenafil was then given in addition to bosentan. After 2 months of combination antipulmonary hypertension therapy, the mean PA pressure was 25 mmHg, the mean capillary pressure was 14 mmHg and the PVR was 2.4 Wood units, allowing listing for heart transplantation alone. Heart transplantation was performed 6 months later with success, no right ventricular dysfunction occurred and antipulmonary hypertension therapy could be weaned without any problem 1 month later.

Cyanosis
Long-term cyanosis increases the postoperative risk for several reasons: systemic-to-pulmonary artery collateral vessels are frequent, often multiple and diffuse, particularly around BTT shunts (the risk of uncontrollable bleeding during and after surgery is then very important, and can contraindicate heart transplantation); long-term cyanosis leads to renal insufficiency, which is susceptible to increase after transplantation, requiring haemodialysis [22], and leads to the opportunity for combined heart–kidney transplantation to be considered; and cyanosis increases susceptibility to infections, which can be particularly deleterious under immunsuppressive therapy.

Pulmonary arteriovenous fistulae, as a well-known complication of partial cavopulmonary connections, such as termino-terminal Glenn anastomosis, can constitute an additional cause of cyanosis. The largest fistulae are eligible for percutaneous closure techniques, but the smaller ones persist and prevent complete relief of cyanosis. The future of these fistulae after heart transplantation is hazardous: they can either disappear or persist, leaving a persistent cyanosis, responsible for higher postoperative mortality and morbidity.

Previous surgery
Most patients with ACHD awaiting heart transplantation have already had cardiac palliative or corrective surgery, often with multiple interventions. In the study by Cohen et al., this proportion reached 90%, with a mean of 2.4 previous operations and a majority (75%) of complex CHDs [6]. An additional procedure during transplantation was necessary in 21.6% of patients in this series (suppression of a BTT shunt, take-down of a partial cavopulmonary connection), and in 30% in the series reported by Irving et al. [23]. This underlines the need to plan surgery carefully to minimize ischaemic time. During donor-heart procurement, extended harvesting of pulmonary arteries, aorta or systemic veins can be necessary to avoid the use of prosthetic material to reconstruct vascular anatomy in the recipient [14]. Difficulties with the surgical approach must be anticipated, and surgical dissection must be done very carefully to avoid injury to cardiac chambers and/or great vessels. In addition, it can be useful to expose the patient’s femoral vessels, to be ready for emergency femoro-femoral cardiopulmonary bypass if needed. All these predictable technical difficulties increase the risk of postoperative bleeding, and account for the high rate of reoperations for surgical haemostasis (24.7% in the series of Cohen et al.) [6]. Preoperative evaluation must take this risk into account, and will include complete right and left heart catheterization and angiography, including a search for aortopulmonary collateral arteries, evaluation of arterial and venous accesses and Doppler evaluation of the venous and arterial pathways. Indeed, previous CHD surgery and previous heart catheterizations are often responsible for thrombosis of central veins, which may complicate the operative course at the time of heart transplantation. Previous surgeries are also responsible for mediastinal adhesions, with, consequently, increased ischaemic times, increased risk of injury to heart or vessels at chest opening and surgical bleeding. Preoperative evaluation will be completed by a computed tomography scan to determine precisely the relationships between the heart, the great vessels and the sternum.

Interventional catheterization procedures can be extremely useful in this context, to reduce ischaemic time during transplantation. Device embolization of aortopulmonary shunts (BTT) or occlusion of a Potts anastomosis by a covered stent in the descending aorta can be performed immediately before heart transplantation, if the occlusion test during pretransplant catheterization was favourable [24]. Aortopulmonary collateral vessels, evaluated by cardiac angiography and/or computed tomography angiography, can be embolized before transplantation with coils or other devices, but the results of these procedures are often incomplete, and collateral circulation remains a major risk factor for immediate postoperative mortality and bleeding [15]. For this reason, multiple aortopulmonary collaterals can contraindicate heart transplantation in certain patients.

Scoliosis is frequently associated with cyanotic congenital heart defects and previous surgery via thoracotomy (aortopulmonary shunts, repair of aortic coarctation). Scoliosis can lead to chronic respiratory insufficiency, which, if severe, can be a contraindication to heart transplantation. It is important to note that the presence of a former thoracotomy, which is frequent in patients with complex CHD, is an absolute contraindication to heart–lung transplantation because of the major risk of perioperative uncontrollable bleeding.

Allosensitization: development of anti-human leucocyte antigen antibodies
Patients with ACHD are particularly exposed to human leucocyte antigen (HLA) allosensitization because of previous surgeries, especially those requiring cardiac bypass, which is responsible for multiple blood transfusions [25]. The use of human homograft material is also a source of immunization, with subsequent development of circulating anti-HLA antibodies [25]. The presence of anti-HLA antibodies limits the number of potentially compatible donors, as many teams avoid transplanting in the presence of "forbidden antigens" (positive virtual cross-match) because of the risk of acute rejection, particularly humoral or antibody-mediated, and reduced long-term survival because of chronic rejection and graft vasculopathy [26]. The time on the waiting list is therefore longer for sensitized patients, and mortality during waiting time is increased. Many protocols have been used to reduce HLA allosensitization before transplantation (repeated plasmapheresis, immunoglobulins, rituximab, etc.), but their results are uncertain.
Desensitization by plasmapheresis is a long and demanding treatment, and entails a high risk of sudden cardiac failure and circulatory arrest in patients with precarious haemodynamics awaiting transplantation. Treatment with rituximab leads to an increased risk of infection [25]. Moreover, interpretation of anti-HLA antibody screening is sometimes difficult, because of the increasing sensitivity of the screening techniques, and has to be balanced with the uncertainties concerning the clinical impact of these antibodies.

Failure of other organs

Kidney
Severe chronic renal insufficiency is a contraindication to isolated heart transplantation, and combined heart and kidney transplantation should then be considered. Moreover, renal insufficiency, even if moderate (creatininaemia > 1.5 mg/dL [132 μmol/L]) is a strong predictor of mortality after heart transplantation [27].

Liver
Chronic hepatic failure is also a strong predictor of mortality after heart transplantation. Some patients, especially those with TGA operated on by the atrial switch technique, can develop hepatic insufficiency and cirrhosis when the systemic venous pathway is stenotic or occluded. Fontan patients can also develop liver cirrhosis. Preoperative evaluation should include hepatic evaluation, including hepatic biopsy to grade the degree of cirrhosis and hepatic fibrosis. An increased or modified Model for End-stage Liver Disease (MELD) score is a predictor of mortality and morbidity after heart transplantation [28]. Combined heart–liver transplantation can be considered in some cases.

Transplantation with physiological single lung

Adults with complex CHD can have a physiological single lung, either anatomically (tetralogy of Fallot with or without pulmonary atresia, with absence or early occlusion of a PA [usually the left PA because of arterial duct closure in infancy]) or functionally (unilateral pulmonary hypertension secondary to previous aortopulmonary shunts or collateral arteries or — less frequently — pulmonary venous obstruction). Heart transplantation “on a single lung” can be performed in these cases, with acceptable results, as demonstrated in a recent retrospective study of 22 patients [29]. In this study, no difference in survival was found between single lung and other heart transplant patients. The authors advocate aggressive pretransplant occlusion of collateral vessels by interventional catheterization procedures, to reduce postoperative risk [29].

Failing Fontan: specific problems

Heart transplantation in patients with a functionally univentricular heart palliated by Fontan-type surgery raises some specific and difficult problems. These patients have always undergone one or more previous cardiac surgeries. The physiology of the failing Fontan leads to a variety of clinical presentations: congestive heart failure because of ventricular dysfunction, severe atrioventricular valve insufficiency and arrhythmias, low cardiac output, ascites, chronic oedema, cyanosis, and renal or hepatic insufficiency. Protein-losing enteropathy (PLE) and its variant — plastic bronchitis — are specific to failing Fontan circulation, and heart transplantation is now recognized as the only valuable therapeutic tool in these cases.

PLE is defined by an enteral protein loss, leading to hypoproteinaemia and hypoalbuminaemia. Patients with long-standing chronic PLE have nutritional impairment, with vitamin and immune deficiencies. These characteristics can explain why, although heart transplantation seems currently to be the most effective treatment for PLE, with resolution rates varying from to 70% to 100% [30], post-transplant management is often difficult, and some reports describe higher rates of complications after heart transplantation (infections, graft failure and increased mortality) [31]. A recent multicentre study, comparing the results of heart transplantation in 70 Fontan patients with PLE and 173 without PLE, was more optimistic, finding no association of PLE with rejection, infection or death [32]. However, PLE patients in this study were not stratified by disease severity, and serum protein and albumin concentrations were not different between the two groups of patients, indicating that PLE was rather moderate. From our limited experience, we think that PLE patients should be put on the waiting list relatively early, before ionic, vitamin, iron and albumin loss become too important. Other publications support these conclusions, and extend the recommendation of earlier referral for heart transplantation to all failing Fontan patients, before multiorgan failure [30]. Failing Fontan patients with PLE and preserved ventricular function were deemed to have higher early and late mortality than those with impaired ventricular function [31], but this finding has since been challenged [30].

In our institution, we have transplanted three patients with PLE, two of whom had complete resolution. The third patient had end-stage PLE, with extreme alteration of blood tests; after initial improvement, he experienced recurrence of severe PLE, and died 6 months later of acute lymphoma. This fatal evolution may have been favoured by the immune deficiency caused by end-stage PLE, coupled with the necessary immunosuppressive treatment. A lower immunosuppressive regimen has been recommended in this population to minimize the risk of infections or cancer [28].

Previous Fontan-type operations are a source of technical problems for the surgeon, with the need to reconstruct the caval veins and often the PAs, which can require extended vascular harvesting of the superior caval vein, innominate vein and PAs of the donor [30]. Previous Fontan conversion to extracardiac conduit does not seem to increase the operative risk [30].

Postoperative and long-term outcomes

Two recent studies confirmed that perioperative mortality and morbidity are higher in ACHD recipients than in patients transplanted for other causes [4,5]. The first study analyzed a cohort of 737 patients with ACHD of 26993 orthotopic heart transplant patients from the UNOS registry in the USA [5]. In this series, ACHD patients were younger, were more likely female, had fewer ventricular assist devices, were more likely to have class II anti-HLA antibodies and were listed for a longer time (249 vs 181 days; P < 0.01). The operative mortality was higher (11.5% vs 4.6%; P < 0.001). Patients with ACHD had a longer postoperative length of stay (27 vs 20
days; \(P < 0.01\), a higher incidence of primary graft dysfunction (4.3% vs 2.6%; \(P < 0.01\)) and a higher need for dialysis (20% vs 9%; \(P < 0.01\)). Primary graft dysfunction was the most common cause of death (5.8%). However, ACHD recipients who survive the first post-transplant year have better long-term survival than other recipients [5]. The second study, a meta-analysis [4], confirmed that 30-day mortality was higher in ACHD heart transplant recipients (risk ratio 2.18). However, 10-year mortality was significantly lower in these patients (risk ratio 0.75). Whereas death secondary to primary graft failure, stroke and haemorrhage was significantly higher in ACHD patients, death caused by malignancy, infection, rejection or cardiac allograft vasculopathy was decreased, although only death from malignancy achieved significance [4]. This latter finding is probably a reflection of the combined effects of younger age and absence of risk factors for coronary disease.

Heart–lung and lung transplantation

Eisenmenger syndrome

Indication and timing for inscription on the waiting list are particularly challenging in patients with Eisenmenger syndrome, because of their often-prolonged spontaneous survival, which may be improved by antihypertensive therapies [33]. This survival rate must be balanced with the high rate of complications after heart–lung transplantation [34]. Heart–lung transplantation is generally indicated for end-stage Eisenmenger disease on irreparable CHD, with dysfunction of one or both ventricles. The level of PVR is not part of the indication process [35]. Few series are available in the literature. A recent analysis of a cohort of 50 patients transplanted for pulmonary arterial hypertension associated with CHD in our institution (1987–2016) reported a survival rate of 75% at 1 year, 60% at 5 years and 50% at 10 years (M. Pontaillier, unpublished data). In this series, risk factors for mortality on the waiting list were right ventricular dysfunction and elevated right atrial pressures, and the risk factor for operative mortality was New York Heart Association stage IV; for both, antihypertensive therapy was a protective factor (M. Pontaillier, unpublished data). Similar results were published by a German team in 2007 (80% survival at 1 year, 69% at 5 years and 53% at 10 years), with a tendency towards better long-time survival of ACHD patients compared with heart–lung and double lung transplantation in other patients [36]. However, because of the lack of donors, heart–lung transplantation is now increasingly abandoned, when possible, in favour of double-lung transplantation with intracardiac repair [35]. In our centre, two young women with Eisenmenger syndrome caused by an ostium secundum atrial septal defect underwent double-lung transplantation, then percutaneous closure of the atrial septal defect with an AMPLATZER® device (St. Jude Medical, St. Paul, MN, USA) a few months later, with an excellent outcome.

Tetralogy of Fallot with pulmonary atresia

These patients are often contraindicated for heart transplantation because of the small size of the pulmonary branches, with possible localized pulmonary hypertension. However, collateral circulation and previous aortopulmonary shunts, with thoracotomy scars, often preclude heart–lung transplantation as well.

Mechanical circulatory support and ventricular assist devices

The annual report of the French "Agence de Biomédecine" indicated that in 2015, 25% of adult heart transplant recipients were on mechanical circulatory support at the time of transplantation. However, this is not the case for patients with ACHD, because the complexity of the anatomy and previous interventions often preclude the use of this type of support. A recent American Heart Association statement confirmed this tendency, and pointed out the disadvantage of patients with ACHD on the waiting list compared with adults with acquired heart failure [37]. When clinical status of the patient warrants the use of mechanical circulatory support, careful evaluation must be performed with the surgeon, especially regarding the anatomy and permeability of the great vessels. Extracorporeal membrane oxygenation is the simplest type of mechanical circulatory support, but is meant to last only a few weeks; its use as a bridge for transplantation is associated with decreased in-hospital survival in patients with CHD (44% for CHD with two-ventricle physiology and 33% in CHD with univentricular physiology compared with 63% for cardiomyopathy) [38]. Ventricular assist devices are difficult to use in ACHD, especially in patients with univentricular hearts and Fontan-type circulation [39]. Moreover, biventricular support is often needed in ACHD. The EXCOR® biventricular device (Berlin Heart, Berlin, Germany) has been used with success in a few failing Fontan patients as a bridge-to-transplant, as well as HeartMate II (Thoratec, Pleasanton, CA, USA) and HeartWare® (HeartWare, Framingham, MA, USA) [37]. As in patients without CHD, complications are common with ventricular assist devices, especially stroke, followed by infection, bleeding and thromboembolic events [40].

Conclusions

Heart transplantation is the final milestone for many patients with ACHD who develop end-stage heart failure. Early mortality and morbidity remain high because of the particularities of these patients, including technical challenges for the surgeon (frequent previous surgeries and anatomic complexity), the frequent involvement of other organs, such as kidney, liver and lungs, and the physiology of Fontan-type circulation. This has led some authors to recommend that heart transplantation in ACHD should be performed by a surgeon specialized in congenital heart defects, in an adult care environment [41]. However, long-term survival is better in patients with ACHD than in adults without CHD, which should encourage us to continue to refine the indications, the time of listing and pre-, peri- and postoperative care, to reduce perioperative mortality, which remains "the Achilles heel" of heart transplantation in ACHD [5].
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