Surgical treatment concepts for end-stage congenital heart diseases

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ABSTRACT

It is anticipated that as many as 10-20% of patients alive with anatomical congenital heart lesions may eventually develop heart failure. Most of these patients have undergone previous palliative or corrective surgeries. The Fontan procedure, although it has helped many patients with single-ventricle physiology to become hemodynamically functional, it is associated with protein-losing entopathy, intractable atrial arrhythmia and systemic ventricular dysfunction. In patients with transposition of the great arteries, physiological or intra-atrial repair techniques (Mustard and Senning) or anatomical correction (arterial switch) have been used. The majority of these patients are clinically well throughout their adult life but, as a consequence of the potential for right ventricular failure, some may develop cardiac insufficiency. Although long-term survival and quality of life in children and adults with complex congenital heart disease have remarkably improved due to advances in operative techniques and perioperative management, as well as the increasing experience of congenital heart surgeons, a growing number of these patients eventually develop end-stage heart failure and will require another treatment. Surgical options for treatment of these patients are limited. Heart transplantation has become a well-established treatment option for children as well as adults with end-stage congenital heart disease. Because of organ donor shortage, another option is mechanical circulatory assist device implantation, either as a bridge to transplantation, or as a permanent therapy.

Keywords: end-stage heart failure, single ventricle physiology, atrial switch, heart transplantation, mechanical circulatory assist devices.

INTRODUCTION

Great developments and progress in cardiac surgery and cardiology have resulted in pediatric patients with congenital heart disease (CHD) who previously would have died now reaching adulthood. Although long-term survival and quality of life in children and adults with complex congenital heart disease have remarkably improved due to advances in operative techniques and perioperative management, as well as the increasing experience of congenital heart surgeons, a growing number of patients with complex CHD eventually develop end-stage heart failure and will require another treatment. These may be the patients with single ventricle physiology and failed Fontan circulation, those with transposition of the great arteries, either palliated or corrected, and those with situs inversus. What do we have to offer them then? Two alternatives remain: heart transplantation or permanent mechanical circulatory support.
Single-ventricle physiology

The Fontan procedure, initially conceived for a patient with tricuspid atresia (1) has become established as a definitive palliation for cardiac anomalies with a single functional ventricle or two ventricles with anatomy unsuitable for biventricular repair. However, intermediate and long-term results showed that the Fontan operation was associated with a high incidence of supraventricular arrhythmia, circulatory pathway obstruction, decreased ventricular function, increasing cyanosis, and protein-losing enteropathy (PLE). This led to Fontan revision procedures.

Total cavopulmonary artery connection was introduced to reduce the incidence of supraventricular arrhythmia, by construction of a cavopulmonary anastomosis for the superior vena cava (Glenn procedure) and either an intra-atrial tunnel or an extracardiac conduit for the drainage of the inferior vena cava into the pulmonary artery. Still a considerable number of patients experienced late failure. PLE is a life-threatening complication after the Fontan procedure, the prevalence of which among the 30-day survivors ranges from 0-25%. It has a very dismal prognosis, with either medical or surgical treatment, with a reported mortality of 46-64% (2, 3). The issue of failing Fontan has then been an ever increasing burden to univentricular patients.

Transposition of the great arteries

In patients with transposition of the great arteries (TGA), physiological or intra-atrial repair techniques (Mustard and Senning) have been used for more than three decades. Given that the Senning procedure can be performed without having to use prosthetic material for redirection of the blood flow, this was the most frequently used technique, considering its potential to allow growth. Anatomical correction (arterial switch) avoids the use of the left ventricle as a systemic ventricle, but as it was developed later on, in the 1970s, there are still many patients who reach adulthood with some variation of the intra-atrial repair technique. The majority of these patients are clinically well throughout their adult life but, as a consequence of the potential for right ventricular (systemic) failure, some may develop cardiac insufficiency. Surgical options for treatment of these patients are limited. Substitution of the tricuspid valve is generally insufficient to avoid functional worsening. Another option would be late conversion to a two-stage arterial switch: banding of the pulmonary artery and later take-down of the intra-atrial repair and arterial switch. At first this technique presented encouraging results. Later studies, with longer follow-up time, confirmed that the success rate was lower than 20% in patients older than 12 years of age. The main cause of failure was the inability of the LV to adapt to systemic pressure. Given that the majority of patients who develop ventricular failure are in their 20s or 30s, this technique is not considered a true alternative.

Faced with this situation, for patients who progress toward cardiac insufficiency, heart transplantation is an interesting therapeutic option. The poor results with other surgical techniques, such as the late arterial switch, make heart transplantation a good therapeutic alternative in cases where right ventricular insufficiency develops.

Our experience in transplantation for end-stage congenital heart diseases

Patients with complex CHD, either previously palliated or remaining untreated, present anatomical, physiological and technical challenges to the transplant surgeon (4, 5). Specific anatomical abnormalities such as vascular and cardiac size, position...
and situs necessitate modifications of each component (6, 7). These technical dilemmas have been overcome by the continuously evolving surgical ingenuity and creativity born out of practice and experience, innovative solutions and careful surgical planning, adapting the complex recipient anatomy to the normal donor anatomy. Situs inversus requires a spatial arrangement of the systemic venous drainage (6, 7) and, despite its anatomical and technical complexities, this did not preclude our group from performing heart transplantation by anastomosing the anatomic constants such as the left atrium, pulmonary artery and aorta in the usual manner, and by in situ systemic venous reconstruction. The reconstructed venous passageways have remained patent and unobstructed for more than 12 years after the operation. We were also able to carry out heart transplantation in 13 children (age range 1.6-17 years) with transposition of the great arteries palliated with atrial switch (Mustard/Senning) procedure, in 2 children (1 and 6 months old) who had a previous arterial switch operation and in 6 patients (16-59 years old) with congenitally corrected transposition of the great arteries, who all had end-stage cardiac failure. Because of several previous surgical interventions and the orientation of the great vessels, the operation presented some technical problems. The operative procedures and implantation of the donor heart were modified accordingly. The technical results were satisfactory and now they (longest survivor 16 years: range 6-16 years post-transplantation) are doing better than ever before in their lives. Likewise, heart transplantation was done on univentricular hearts with failed Fontan circulation in 10 patients (5 children, age range 2-16 years; 5 adults, age range 21-58 years). The procedure was technically elaborate but not difficult to perform, similar to the experience of Kanter et al (8) who stated that despite its being a more complex transplant operation they did not find any early or midterm disadvantage for the 27 children who underwent heart transplantation after a previous Fontan procedure. We contend that carefully selected children with a failing Fontan circulation can do as well as other children with heart transplantation; however, we noted that this population has increased perioperative morbidity (perioperative hemorrhage due to previous repeated sternotomies) and 40% mortality due to increased pulmonary vascular resistance, wherein the pulmonary vascular changes which developed over time were difficult to identify. Mitchell et al. (9) emphasized the importance of avoiding donors with marginal cardiac functions or with excessive inotrope requirements, as well as donor-to-recipient weight ratios of <1:1.

Michielon and his group (10) made the same recommendations and pointed out that successful outcome is primarily related to the number of desirable donor criteria and donor-recipient matching. This tackles the issue of acceptance of less-than-ideal donor hearts under the pressure of deteriorating clinical status of the recipient. Because of donor shortage, do we then have a choice? Options could be to treat individual Fontan circulation problems, i.e. add anti-arrhythmia surgery (11, 12) when applicable, early relief of any anatomical obstruction, performing bidirectional Glenn shunt while listing the patient for heart transplantation, or proceed directly to transplantation rather than Fontan revision. Presently, there are no reports about proven benefits of these options, if they have ever been tried. It is unlikely that there will be enough donor hearts for these patients if or when the Fontan circulations fail in the late postoperative period. Perhaps the best hope for this growing population lies in the developments in implantable devic-
es rather than in transplantation. For the patient with a single ventricle, mechanical support may be very difficult, although not impossible.

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