Archives of Cardiovascular Disease (2017) xxx, xxx—xxx

Available online at ScienceDirect www.sciencedirect.com

Elsevier Masson France EM|consulte www.em-consulte.com/en

REVIEW

Heart transplantation in adults with congenital heart disease

Transplantation cardiaque chez les congénitaux adultes

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Received 29 November 2016; received in revised form 4 January 2017; accepted 4 January 2017

KEYWORDS
Adult congenital heart disease;
Heart transplantation;
Heart—lung transplantation;
Pulmonary vascular resistance

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

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; VO_{2max}, maximal oxygen uptake.

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http://dx.doi.org/10.1016/j.acvd.2017.01.002

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.

MOTS CLÉS
Cardiopathies congénitales de l'adulte ;
Transplantation cardiaque ;
Transplantation cardiopulmonaire ;
Résistances vasculaires pulmonaires

Résumé Avec les progrès de la chirurgie cardiaque congénitale et de la prise en charge postopératoire, un nombre croissant d'enfants avec des cardiopathies congénitales complexes atteignent maintenant l'âge adulte. On estime qu'il existe actuellement plus d'adultes que d'enfants vivant avec une cardiopathie congénitale, y compris avec une cardiopathie complexe. Parmi ces adultes congénitaux, un certain nombre vont développer avec le temps une dysfonction ventriculaire. L'insuffisance cardiaque représente actuellement 26—42 % des causes de décès chez les adultes ayant une cardiopathie congénitale, opérée ou non. Pour ces patients, la transplantation cardiaque, ou cardio-pulmonaire en cas de syndrome d'Eisenmenger, représente l'ultime possibilité thérapeutique. Cette population est réputée à haut risque de mortalité post-opératoire immédiate, bien que la survie à long-terme soit en fait comparable à celle des patients transplantés pour d'autres raisons. La transplantation cardiaque représente souvent un défi sur le plan chirurgical et de la prise en charge, en raison de la complexité anatomique et des abords vasculaires, des interventions préalables palliatives ou correctrices, des conséquences du bas débit cardiaque prolongé et/ou de la cyanose sur les autres organes (reins, foie, poumons), avec souvent une élévation des résistances vasculaires pulmonaires. Dans cette revue, nous aborderons les problèmes spécifiques que pose la transplantation cardiaque ou cardiopulmonaire dans cette population, revisiterons les indications et contre-indications, et actualiserons le devenir à long terme et le pronostic.

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

Please cite this article in press as: Houyel L, et al. Heart transplantation in adults with congenital heart disease. Arch Cardiovasc Dis (2017), http://dx.doi.org/10.1016/j.acvd.2017.01.002