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Clinical characteristics and outcome of heart transplant in adults with Congenital Heart Diseases. Subanalysis of Spanish Registry on Heart Transplantation

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INTRODUCTION: In spite of recent medical and surgical advances, some patients with congenital heart diseases (CHD) need a heart transplant (HTx) as adults. Consequently, It has become necessary to know clinical characteristics of these HTx candidates.
OBJETIVE: To analyze outcome in the Spanish adult transplanted population with CHD and compare it with the most frequent causes of adult transplantation (ischemic heart disease [IHD] and idiopathic dilated cardiomyopathy [IDCM]) and among different subgroups of CHD.
MATERIALS AND METHODS: From May 1984 to December 2009, 6,048 patients were transplanted in Spain. Pediatric transplants(<16 years), combined transplants, reHTx, HTx performed in other centers without adult CHD cases, and HTx for heart diseases other than IDCM and IHD were excluded. Therefore, study population included 3.166 patients (IHD: 1 888; IDCM: 1,223; CHD: 55).
Study subgroups: CHD transplants were classified according to pathophysiology into four groups: 1) Single ventricle with different degrees of pulmonary stenosis (n:18). 2) Single ventricle with cavopulmonary shunt surgery (Glenn/Fontan) (n:10). 3) Congenitally corrected transposition of great arteries or transposition of great arteries with Mustard/Senning surgery (n:10). 4) CHDs with different degrees of right ventricle overload (tetralogy of Fallot, Ebstein's disease, transposition of great arteries with pulmonary stenosis and Rastelli patch, double outlet right ventricle) (n:17).
RESULTS: Significant differences were found between the clinical profile of patients with CHD and other groups: younger age, less need for inotropes and minor renal dysfunction. The analysis of survival showed some differences between groups (CHD vs IHD, p=0.05; CHD vs IDCM, p=0.5; IHD vs IDCM, p=0.0001). Early mortality was different between the different CHD subgroups (group 1: 19%, group 2: 40%, group 3: 0%, group 4: 29%; p<0.001). The probability of overall survival was higher in CHD group, despite its high early mortality.
CONCLUSIONS: Percentage of adult with CHD transplanted in Spain is low (1%). Long term survival is higher compared with groups with different indications for HTx (IHD and IDCM). Nevertheless, early mortality is high in operated or unoperated patients with single ventricle physiology. European registries with a larger number of patients are needed to better define results in this group of patients.