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Tricuspid regurgitation or Ebsteinoid dysplasia of the tricuspid valve in congenitally corrected transposition: Is valvuloplasty necessary at anatomic repair?

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Objectives: Patients with congenitally corrected transposition of the great arteries can present with tricuspid regurgitation (TR) and Ebsteinoid dysplasia of the tricuspid valve. To determine the fate of the tricuspid valve after anatomic repair and the effectiveness of tricuspid valvuloplasty, we reviewed our experience with anatomic repair of congenitally corrected transposition of the great arteries.

Methods: From 1992 to 2012, 106 patients with congenitally corrected transposition of the great arteries underwent anatomic repair. Of the 106 patients, 24 (22.6%) had moderate or greater TR before anatomic repair, 14 of whom had Ebsteinoid dysplasia. Nine patients (8.5%) had Ebsteinoid dysplasia without significant TR, and 73 patients (68.9%) had neither TR nor dysplasia.

Results: Of the 106 patients, 6 (5.6%) with TR underwent tricuspid valvuloplasty at anatomic repair, 5 with (21%) and 1 without (11%) Ebsteinoid dysplasia ($P < .001$). During a median follow-up period of 32 months, of the 24 patients with TR before anatomic repair, all 6 who had undergone tricuspid valvuloplasty had mild TR or less at the latest follow-up visit; 15 of the 18 (83%) without valvuloplasty had mild TR or less ($P = .4$) and 3 (16.7%) had moderate or greater TR. Of the 14 patients with Ebsteinoid dysplasia and TR, 5 underwent valvuloplasty and had no significant TR during follow-up; 2 of the 9 (22.2%) without valvuloplasty had moderate or greater TR ($P = .51$). Valvuloplasty was associated with an absolute risk of TR reduction of 16.7%, which was further reduced by 22.2% in patients with associated Ebsteinoid dysplasia.

Conclusions: Tricuspid valve function significantly improved after anatomic repair, independent of direct surgical intervention. For significant TR associated with Ebsteinoid dysplasia, valvuloplasty should be considered. (J Thorac Cardiovasc Surg 2014;147:576-80)

Congenitally corrected transposition of the great arteries (ccTGA), although rare and representing <1% of all congenital heart defects,[1] is often associated with tricuspid regurgitation (TR), with or without dysplasia of the tricuspid valve that mimics the tricuspid valve dysplasia seen in Ebstein's anomaly.[2] TR is an integral part of the natural history of this anatomy owing to functional tricuspid valve dysfunction, because it is associated with right ventricular (RV) dysfunction. It will improve when the left ventricular (LV) and RV pressures are equalized or inversed and the ventricular septum shifts, such as after pulmonary artery banding for LV preparation[3,4] or anatomic repair,[5] placing the tricuspid valve as the low-pressure, subpulmonary atrioventricular valve.

Ebsteinoid dysplasia of the tricuspid valve in ccTGA represents a variable spectrum of disease, with failure of delamination of the tricuspid valve leaflets from the RV wall and subsequent tethering and apical displacement of the leaflet hinge points, maximally at the commissure between the posterior and septal leaflets.[6] We have preferred the term ''Ebsteinoid dysplasia'' for this lesion, because significant differences exist between it and Ebstein's anomaly[7] with a concordant atrioventricular connection compared with the tricuspid dysplasia seen in atrioventricular discordance: The atrialized portion of the right ventricle is not thinned, rarely the mural or septal leaflet can be absent, and it has been associated with a ventricular septal defect[6] and aortic arch obstruction.[8]

Physiologic repair of ccTGA, in which the right ventricle is left in the systemic position, and associated anomalies, such as TR, ventricular septal defect, and pulmonary atresia, are repaired, carries a high risk, and patients are often referred late for surgical repair, when the systemic RV function has started to decline.[9] In this setting, repair

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Abbreviations and Acronyms

ccTGA = Congenitally corrected transposition of the great arteries
LV = left ventricular
mRV = morphologic right ventricle
RV = right ventricular
TR = tricuspid regurgitation

has provided poor results, given that the tricuspid valve supports the systemic circulation, and valve replacement has been advocated,[10] albeit at a high operative risk. Anatomic repair, which corrects atrioventricular and ventriculoarterial discordance using a double-switch procedure, places the right ventricle and tricuspid valve in the pulmonary position. It is a promising solution that has shown low operative mortality and good functional results compared with physiologic repair.[1,11-14]

Murtuza and colleagues[14] reported, among 113 patients who had undergone anatomic repair, a prevalence of Ebsteinoid dysplasia of the tricuspid valve of 12.3% (n = 14), and 6 patients (5.3%) had severe TR. Currently, little information is available regarding the best management of significant TR, in particular, when associated with Ebsteinoid dysplasia at anatomic repair of ccTGA. The goal of the present study was to review our experience in managing the tricuspid valve in patients with significant TR at anatomic repair and infer the best surgical option for treatment.

METHODS

Study Design

The present study was a retrospective review of all patients with ccTGA who had undergone anatomic repair at our institution from 1992 to 2012. The Boston Children's Hospital institutional review board approved the present study, and individual patient consent was waived.

The primary endpoints were moderate or greater TR and tricuspid valve reoperation after anatomic repair. The clinical and treatment variables were recorded to determine the predictors of the endpoints. All patients underwent follow-up examination to death or June 2012.

Surgical Technique

The surgical techniques used in the present study population have been previously reported in detail.[15] The decision to address the tricuspid valve at anatomic repair was determined from the preoperative echocardiographic findings of the presence of moderate or greater TR and a discernable structural defect, such as a tethered septal leaflet, cleft, or splayed-open anteroseptal commissure. The surgical techniques were tailored to the mechanism responsible for regurgitation at the discretion of the operating surgeon. Exposing the tricuspid valve can be challenging in patients with ccTGA and mesocardia, because the ventricular mass lies anterior to the atrial mass. In these patients, we rotate the ventricular mass into the left pleural space in situs solitus or the right pleural space in situs inversus to expose the tricuspid valve.

Statistical Analysis

Statistical analyses were performed using the Statistical Package for Social Sciences software, version 21 (SPSS, Inc, Chicago, Ill). The data are presented as mean ± standard deviation or median and range, as appropriate. Continuous variables were analyzed using the related samples Wilcoxon signed rank test or the Kruskal-Wallis 1-way analysis of variance test, and categorical variables using the chi-square test or Fisher's exact test. All statistical tests were 2-tailed, and $P < .05$ was considered significant.

RESULTS

Study Population

The study population has previously been described in detail.[16] A total of 106 consecutive patients underwent anatomic repair for ccTGA during the study period. The median age at surgery was 1.2 years (range, 2 months to 43 years). The segmental anatomy was SLL (situs solitus, L-loop ventricles, L-transposed great arteries) in 93 patients (87.7%) and IDD (situs inversus, D-loop ventricules, D-transposed great arteries) in 13 patients (12.3%). Levocardia was present in 63 patients (59.4%), dextrocardia in 24 (22.6%), and mesocardia in 9 (8.5%). Atrial switch was accomplished using a modified Senning procedure in 35 patients (33%) and a Mustard procedure in 71 (67%). Also, 42 patients (39.6%) had undergone a Rastelli procedure for LV outflow reconstruction and 62 (58.5%) arterial switch for restoration of the ventriculoarterial concordance. Two patients (1.9%) had undergone a Nikaidoh procedure late in our experience.

A detailed breakdown of the patient subgoups and outcomes are summarized in Figure 1. Of the 106 patients, 24 (22.6%) had moderate or severe TR before anatomic repair, 15 of whom had Ebsteinoid dysplasia of the tricuspid valve. Another 9 patients (8.5%) had Ebsteinoid dysplasia without significant TR, and 73 patients (68.9%) had neither TR nor dysplasia and were used as controls. Of the patients with TR, 9 (37.5%) had previously undergone pulmonary artery banding, and 15 of the controls had also (20.5%; $P = .08$). The patient baseline characteristics are summarized in Table 1. More patients with significant TR and/or Ebsteinoid dysplasia had undergone arterial switch (29 of 35) than a Rastelli procedure (6 of 35; $P = .007$).

Tricuspid Valvuloplasty at Anatomic Repair

Six patients (5.6%, all in the TR group) had undergone tricuspid valvuloplasty at anatomic repair, 5 with Ebsteinoid dysplasia (20.8%) and 1 without Ebsteinoid dysplasia (11.1%; $P < .001$). The repair techniques consisted predominantly of anteroseptal commissure closure and suture annuloplasty (Table 2). The predominant lesion responsible for regurgitation found on tricuspid valve inspection was a cleft on the septal leaflet or a splayed-open anteroseptal commissure, with thickened and rolled edges from TR. This was addressed by cleft closure, closure of the regurgitant anteroseptal commissure and/or partial suture annuloplasty, annulus plication, or commissuroplasty. All 6 patients who had undergone valvuloplasty