Cone reconstruction of the tricuspid valve in Ebstein anomaly with or without one and a half ventricle repair

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Objective: We sought to review our institutional experience and midterm results with cone reconstruction in Ebstein anomaly with or without one and a half ventricle repair to evaluate these 2 important surgical methods for the treatment of Ebstein anomaly.

Methods: From January 1, 2004, to October 1, 2009, 30 consecutive patients with Ebstein anomaly underwent cone reconstruction. All patients underwent cone reconstruction, and 20 patients with severe Ebstein anomaly had a bidirectional cavopulmonary shunt constructed. The median age was 60 months (range, 2–192 months). Our criteria to define severe Ebstein anomaly include a severely enlarged right-sided chamber, significant abnormality of the septal leaflet of the tricuspid valve, and hemodynamic instability after cardiopulmonary bypass. Clinical or echocardiographic characteristics were studied both preoperatively and postoperatively.

Results: There was 1 (3.3%) hospital death. Before the operation, tricuspid incompetence was moderate in 8 and severe in 22 patients. Postoperative early echocardiographic analysis showed that tricuspid incompetence was mild in 26 patients, moderate in 3 patients, and severe in 1 patient. After a median follow-up time of 22 months, tricuspid incompetence of 20 patients with bidirectional cavopulmonary shunt was mild in 15 and moderate in 4. Tricuspid incompetence of 10 patients without a bidirectional cavopulmonary shunt from the latest echocardiogram (median follow-up time, 32 months) was mild in 4, moderate in 4, and severe in 2 patients. For patients whose postoperative tricuspid incompetence was beyond mild among the 10 patients without a bidirectional cavopulmonary shunt, preoperative echocardiographic analysis shows a severely dilated right ventricle.

Conclusions: Addition of a bidirectional cavopulmonary shunt to cone reconstruction of the tricuspid valve should be considered for young patients with severe Ebstein anomaly who are at high risk of right ventricular failure after the operation. (*J Thorac Cardiovasc Surg* 2011;141:1178-83)

Since its first description by da Silva and coworkers in 2004,¹ cone reconstruction of the tricuspid valve (TV) is more and more used in the surgical treatment for Ebstein anomaly. The typical cone reconstruction procedure includes complete mobilization of the TV, clockwise rotation of the inferior leaflet to approximate with the proximal edge of the septal leaflet, which completes the cone reconstruction; thinned longitudinal internal plication of the atrialized right ventricle; and plication of the inferior annulus.² Although there have been many methods of tricuspid repair for Ebstein anomaly since the first report by Hunter and Lillehei,³ cone reconstruction is regarded as the closest

anatomic repair method in numerous centers for its production of central blood flow and full coaptation of the tricuspid leaflets.

However, there still remain 2 major concerns about the results of cone reconstruction. One is that there are no long-term or midterm studies to report on whether cone reconstruction could avoid pediatric TV replacement in the young population with Ebstein anomaly who undergo this procedure. The other is that cone reconstruction might deteriorate right ventricular (RV) dysfunction because it decreases the tricuspid annular size and distorts or compresses the main right coronary artery because of the plication in the procedure.

Bidirectional cavopulmonary shunt (BCPS) or one and a half ventricle repair has been used for operations on patients with severe anatomic and functional Ebstein anomaly. In patients with Ebstein anomaly, use of the BCPS procedure might reduce RV preload⁴ and provide preload to the left ventricle, which improves biventricular function.⁵ In addition, the RV unloading permits the borderline tricuspid annular size caused by cone reconstruction and might avoid the need for prosthetic valve replacement in the long term.⁶

The purpose of this report is to review our institutional experiences with cone reconstruction of the TV in Ebstein anomaly with or without one and a half ventricle repair.

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Disclosures: Authors have nothing to disclose with regard to commercial support.

Read at the 90th Annual Meeting of The American Association for Thoracic Surgery, Toronto, Ontario, Canada, May 1–5, 2010.

Received for publication May 3, 2010; revisions received Dec 17, 2010; accepted for publication Jan 7, 2011; available ahead of print Feb 14, 2011.

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0022-5223/\$36.00

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doi:10.1016/j.jtcvs.2011.01.015

Abbreviations and Acronyms

BCPS	= bidirectional cavopulmonary shunt
LV	= left ventricular
NYHA	= New York Heart Association
RV	= right ventricular
TR	= tricuspid regurgitation
TV	= tricuspid valve

MATERIALS AND METHODS

This study was approved by the Animal Welfare and Human Studies Committee at Shanghai Jiaotong University School of Medicine.

Patients' Characteristics

From January 1, 2004, to October 1, 2009, 30 consecutive patients with Ebstein anomaly underwent cone reconstruction of the TV. The patients' demographic parameters are detailed in Table 1. Preoperative and postoperative echocardiograms were analyzed in all patients. The echocardiographic analysis included parameters of (1) the degree of apical displacement of the tricuspid annulus, (2) the severity of tricuspid regurgitation (TR), (3) the degree of anterior tricuspid leaflet mobility, (4) left ventricular (LV) function, (5) RV function, and (6) any atrial level shunts. TR was classified on a scale of trace, mild, moderate, and severe.

All patients were followed, ranging from 4 to 61 months (median follow-up time, 22.5 months). The follow-up information for all patients was obtained from outpatient records. Clinical presentation, New York Heart Association (NYHA) functional class, degree of cyanosis, and grade of TR were assessed at postoperative follow-up.

Statistical Analysis

Data were presented as medians with ranges or means and standard deviations. Statistical analysis was performed with SPSS 13.0 software (SPSS, Inc, Chicago, Ill). The differences between 2 groups were evaluated by using Student's *t* test. TR grade and NYHA functional class were analyzed by using the Cochran–Mantel–Haenszel test.

Surgical Technique

Standard surgical technique was performed through a median sternotomy. After initiation of cardiopulmonary bypass with mild moderate hypothermia, the aorta was crossclamped, and cardioplegic arrest was induced. The right atrium was opened parallel to the atrioventricular groove, and the TV was inspected. Typical cone reconstruction of the TV was used in all patients, as described previously,² and consisted of the following. First, we mobilized the tricuspid leaflets completely by incising all abnormal fibrous and muscular attachments between the body of the leaflet and the RV myocardium. Second, we rotated the inferior leaflet clockwise, suturing to the proximal edge of the septal leaflet, which constructed the cone-shaped valve. Third, we longitudinally internally plicated the atrialized right ventricle to exclude its thin part. Fourth, we plicated the true tricuspid annulus to make up the new annulus at the anatomically correct level. Finally, we anchored the leaflets to the new tricuspid annulus.

In addition to the above routine procedures of typical cone reconstruction, our institutional experiences with cone reconstruction included the following. First, we referred to the size of the mitral annulus as the size of the new tricuspid annulus after reconstruction. Second, in this series 7 patients had significant abnormality of the septal leaflet. For these patients, fresh autologous pericardium was used to enlarge a new septal leaflet of the TV, which was then sutured to the inferior leaflet of the TV. Third, we sutured the septal leaflet into the new annulus in an interrupted manner, sutur-

ing the rest of leaflets continuously. Fourth, we partially longitudinally plicated the atrialized right ventricle in all patients with less than 3 separate 2–0 or 3–0 polypropylene interrupted sutures. For patients with a BCPS, we normally did only 1 separate interrupted suture. Fifth, we performed BCPS in more than 60% of patients with Ebstein anomaly in our center. Finally, we closed all atrial septal defects or cases of patent foramen ovale.

BCPS as an adjunctive surgical method for severe Ebstein anomaly was conducted if patients met the following 3 circumstances. (Actually, this is also the standard to define severe Ebstein anomaly.) The first is the presence of severely preoperative enlarged right ventricle based on echocardiographic analysis. Our standard to evaluate a severely dilated right ventricle is that the atrialized right ventricle occupies more than 50% of the right ventricle in the 4-chamber view and that the anterior leaflet causes significant obstruction of the RV outflow tract (Carpentier type C or D).⁷ The second is significant abnormality of the septal leaflet of the TV based on morphologic analysis in the operating room. The third is hemodynamic instability after separation from cardiopulmonary bypass.

RESULTS

Thirty patients underwent cone reconstruction of the TV for Ebstein anomaly during the study period. The median age at the time of the operation was 60 months (range, 2–192 months). Of the 30 patients, 10 underwent cone reconstruction of the TV alone, with a median age of 21 months (range, 2–192 months). Twenty patients underwent cone reconstruction of the TV with a BCPS constructed together, and their age ranged from 14 to 180 months (median, 81.5 months).

The mean aortic crossclamp time was 64 ± 13 minutes, and the mean bypass time was 102 ± 24 minutes. There was 1 (3.3%) hospital death because of severe ventricular arrhythmia as a result of electrolyte disturbances caused by renal failure after the operation. No late deaths occurred. Six patients had permanent first-grade atrioventricular block. There were no delayed sternal closures or mechanical inotropic support.

The TR results of preoperative, early-term postoperative (before discharge), and medium-term postoperative (at the

TABLE 1. Patients' demographic parameters

Variable	Value
Age, mo (median [range])	60 (2–192)
Cone reconstruction without BCPS	21 (2–192)
Cone reconstruction with BCPS	81.5 (14–180)
Indication	
Cyanosis	22 (73%)
Severely dilated RV	21 (70%)
NYHA class III	19 (63%)
Indication for BCPS	
Severely dilated RV	15
Significant abnormal septal leaflet	7
Hemodynamic instability after CPB	2
Follow-up time	22.5 (4–61)
Cone reconstruction without BCPS	32 (4–61)
Cone reconstruction with BCPS	22 (5–38)

BCPS, Bidirectional cavopulmonary shunt; RV, right ventricle; NYHA, New York Heart Association.

TABLE 2. Medium-term postoperative echocardiographic results

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	Preoperative									Early postoperative						Late postoperative								
	TR			LVEF		RVEF		Atrialized RV/RV		TR			LVEF		RVEF		TR			LVEF		RVEF		
	Mild	Moderate	Severe	N	D	N	D	<50%	>50%	Mild	Moderate	Severe	N	D	N	D	Mild	Moderate	Severe	N	D	N	D	
Without BCPS (n = 10)	0	3	7	8	2	9	1	4	6	8	1	1	10	0	7	3	4	4	2	10	0	7	3	
With BCPS (n = 20)	0	5	15	17	3	18	2	5	15	18	2	0	20	0	20	0	15	4	0	19	0	19	0	
P value																		.05						

TR, Tricuspid regurgitation; LVEF, left ventricular ejection fraction; RVEF, right ventricular ejection fraction; RV, right ventricle; BCPS, bidirectional cavopulmonary shunt.

latest follow-up) echocardiograms were depicted in Table 2. Before the operation, tricuspid incompetence was moderate in 8 and severe in 22 patients. Postoperative early echocardiographic analysis showed that tricuspid incompetence was mild in 26 patients, moderate in 3 patients, and severe in 1 patient ($P < .01$ compared with the preoperative echocardiographic results). After a median follow-up time of 22 months, tricuspid incompetence of 20 patients undergoing the BCPS procedure was mild in 15 and moderate in 4 patients. Tricuspid incompetence of 10 patients without a BCPS from the latest echocardiogram (median, 32 months) was mild in 4, moderate in 4, and severe in 2 patients. For patients whose medium-term postoperative tricuspid incompetence was beyond mild among 10 patients without a BCPS, preoperative echocardiographic analysis showed a severely dilated right ventricle.

Five patients had low LV ejection fraction, but all patients had normal LV ejection fraction at the latest follow-up. No echocardiographic evidence of increased velocity across the TV was found at either early-term follow-up or the latest follow-up. All patients had symptomatic improvement and were in NYHA functional class I or II at follow-up. The difference in NYHA functional class assessment between patients with or without a BCPS was not significant at follow-up.

Of the 20 patients undergoing the BCPS procedure, oxygen saturations examined based on pulse oximetry reached 97% to 100% at follow-up. No clinically evident superior vena cava (SVC)-related complications or pulmonary arteriovenous fistula was detected. Four patients reported facial swelling during postoperative follow-up, but this was not problematic.

DISCUSSION

This study reports the youngest series of patients with Ebstein anomaly undergoing cone reconstruction for TV repair. Although early echocardiographic results showed significant improvement in TR, cone reconstruction alone might not produce satisfactory midterm echocardiographic results for TR in Ebstein anomaly with severely dilated right-sided chambers. This is not consistent with the results of the series by da Silva and associates.⁸ The reason for this might be that the patients in this series were much younger

and presented with more severe morphologic abnormality of the TV. It might be also because of different ethnic populations, especially considering more right-sided chamber anomalies in the Asian population. Although patients with Ebstein anomaly have excellent functional outcomes despite postoperative severe TR,⁹ TR aggravates RV dysfunction caused by longstanding TV displacement superimposed on a myopathic ventricle before surgical intervention for Ebstein anomaly.^{10,11} Quinonez and colleagues¹¹ believe it is important to avoid TR of no more than a mild or moderate degree after TV repair for better RV function and favorable RV remodeling. At the same time, pediatric prosthetic TV replacement is certainly problematic because of the subsequent need for reoperation and thromboembolic complications. Taken together, from this study, cone reconstruction alone might not be enough to keep TV function from the need for TV replacement in patients with severe Ebstein anomaly, especially in the young population.

Another concern for cone reconstruction is traditional plication. Longitudinal plication of the atrialized right ventricle and plication of the true tricuspid annulus can result in TV orifice stenosis and distortion or at least compression of the main right coronary artery. Consequently, right-sided chamber function deteriorates, and there is a greater incidence of ventricular arrhythmia after the operation. Malhotra and associates¹² reported partially plicating the atrialized right ventricle closest to the RV apex. Brown and coworkers¹³ reported plication of the atrialized right ventricle selectively to avoid right coronary artery compromise. In this study we used the same maneuver and did not plicate the atrialized portion all the way to the anatomic tricuspid annulus. In this series only 1 patient had ventricular arrhythmias caused by electrolyte disturbances, and there was no occurrence of acute coronary insufficiency.

There remains long-term controversy about whether a biventricular repair is always better than a single-ventricle repair. From the anatomic viewpoint, a biventricular repair is obviously better than a single-ventricle repair. However, the early and late mortality for biventricular repair is higher than that for single-ventricle repair within similar complex anatomy. These conditions show that biventricular repair is not always better than single-ventricle repair from the

functional viewpoint.¹⁴ Based on this understanding, one and a half ventricle repair or BCPS has been alternatively used as a procedure to reach both anatomic and functional balance in patients with a marginal right ventricle.

Ebstein anomaly might be the most common lesion to use BCPS as an adjunctive surgical method according to Van Arsdel and associates' 38-year follow-up of 114 BCPS cases, as well as our institutional experiences.⁵ The addition of the BCPS procedure to the surgical treatment of Ebstein anomaly might decrease RV work⁴ and improve LV function⁵ and thereby might decrease the operative mortality.¹⁵

Another huge advantage of the BCPS procedure is that it might be valid to produce a competent TV in patients with Ebstein anomaly. Marianeschi and coworkers¹⁶ reported that a BCPS might reduce the rate of TV regurgitation and consequently might decrease the incidence of reoperation for aggressive TV intervention. Our results support their findings. In this series tricuspid incompetence of patients with a BCPS is significantly better than that of patients without a BCPS. As mentioned above, cone reconstruction might lead to postoperative TV stenosis because of plication of the true TV annulus. We believe at the same time that the volume unloading from the BCPS procedure might prevent iatrogenic TV stenosis after cone reconstruction of the TV, which is also commented on by Malhotra and coworkers.¹² There are some anatomic circumstances, including the significant abnormality of the septal leaflet, that make cone reconstruction difficult. For these less-than-ideal circumstances, we believe that the BCPS procedure could assist cone reconstruction in production of an ideal postoperative TV.

The indications for the BCPS procedure in patients with Ebstein anomaly remain controversial. Quinonez and colleagues¹¹ described using a BCPS in critically ill patients with severe RV dysfunction or LV dysfunction. Malhotra and coworkers¹² reported that 31 Ebstein anomaly patients with resting cyanosis or effort-induced cyanosis underwent the BCPS procedure. In this study our criteria to use the BCPS procedure as an adjunctive surgical method for severe Ebstein anomaly focused on the TV's competence and RV function after the cone reconstruction. Based on our experience, a preoperatively severely dilated right ventricle (atrialized right ventricle occupies >50% of the right ventricle and the anterior leaflet causes significant RV outflow tract obstruction), outstanding TV morphologic abnormality, and hemodynamic instability after separation from cardiopulmonary bypass are high risk factors for TV reintervention and RV failure. For these patients, no matter whether RV failure or severe cyanosis exists, our surgical strategy is the addition of the BCPS procedure to cone reconstruction of the TV to maintain TR at no more than a moderate degree and to prevent postoperative RV failure.

Of note, the BCPS procedure is associated with SVC syndrome-related complications and collateral veins or pulmonary arteriovenous fistulas. Numata and associates¹⁷ reported that one and a half ventricle repair could not provide more efficient overall circulation and better functional status than Fontan circulation. However, in our series only 4 patients with a BCPS reported facial swelling at follow-up, without problematic symptoms. It is necessary to point out that one and a half ventricle repair has been expanded to many lesions, and different long-term results depend on different lesions. Kim and colleagues⁶ noted in their 38-year follow-up of 114 one and a half ventricle repairs that Ebstein anomaly was associated with the most favorable outcome after one and a half ventricle repair, which is consistent with our experience. We believe that young patients with Ebstein anomaly have the best indication for one and a half ventricle repair. It is a long-term anticipation that one and a half ventricle repair might converse to biventricular repair if the size of the right chamber grows and becomes sufficiently large after the BCPS procedure. We expect some patients with a BCPS in this series might have the chance to convert to biventricular repair in the future.

In conclusion, although cone reconstruction is the most anatomic repair, our results showed that cone reconstruction might not have the expected anatomic result if the patient's condition could not endure anatomic repair. Young patients with severe Ebstein anomaly who are at a high incidence of TV reintervention and a high risk of RV failure after the operation comprise the population in which cone reconstruction might not provide the ideal anatomic effect, and one and a half ventricle repair should be considered as a planned procedure.

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Discussion

Dr Pedro J. del Nido (Boston, Mass). Thank you very much, Professor Liu. I don't think we can match your numbers in total volume.

This is a very important article in that it brings out one of the potential complications to one of the potential long-term problems with using any single procedure for a wide variety of anatomic presentations. Your results are outstanding, and your conclusions that the BCPS adds to the late results is actually quite interesting, although I think somewhat controversial.

First of all, I have a few questions and then a comment. Was there any difference in early mortality or morbidity between the 2 groups, between the group that received only the cone procedure and the group that received the cone procedure plus the BCPS procedure?

Dr Liu. Thank you, Dr del Nido, for your comments and the question.

In this group we compared TV regurgitation after the operation. Before the operation, for the BCPS group, we found severe regurgitation in almost all 20 patients. We had 10 patients with severe regurgitation from this group. However, after the operation, 15 patient had mild regurgitations, and 4 had moderate regurgitation. In the group without a BCPS, we found 4 patients with mild, 4 patients with moderate, and 2 patients with severe regurgitation, and therefore they had different results.

Dr del Nido. I guess that sort of leads me to my next question; that is, this study represents your entire experience, I think, with the cone procedure because you started in 2004. Therefore it includes your learning curve for these patients. As we all know, the learning curve for the cone procedure is quite steep. When did you start doing the BCPS procedure in these patients, and did you start from the beginning deciding which group would have the BCPS procedure, or did you start later in your experience?

Dr Liu. Before we do the cone reconstruction procedure, we also do some for patients with Ebstein anomaly. Since 2004, we have done the cone reconstruction procedure. At the start, we did not do the bidirectional Glenn procedure and then during the follow-up, we found that some things were not so good. Therefore we compared the patients with and without the bidirectional Glenn procedure, and we found that the group with the bidirectional Glenn procedure had good results.

Therefore we now have 3 stages. First, before the operation, based on echocardiographic analysis, if the patient has an enlarged right ventricle and the echocardiogram shows very severe regurgitation of the TV and also during the operation we find very severe regurgitation of the TV, it is also very difficult to separate the patient from cardiopulmonary bypass, and therefore we need to do the BCPS procedure.

Dr del Nido. The other question I had, because your experience is very different than our experience, is that we have been doing the cone procedure not quite as long; I think we have only been doing it for about 3 years. We have more than 40 patients now, and we do not have a single patient in whom we have had to do the Glenn procedure, the bicaval pulmonary shunt, to come off bypass or to preserve RV function. Our late results are that this procedure, the cone procedure alone, is actually quite durable. In intermediate follow-up at about 2 years, we do not see any recurrence of TR.

Therefore this makes me wonder. One of the concerns I have always had about the cone procedure is that there is not much discussion about the more severe cases in which you have not just a very tethered anterior leaflet but one in which the anterior leaflet actually is attached to the septum and therefore you have tricuspid stenosis in addition to severe regurgitation. Have you been able to analyze the patients on whom you operated on and had difficulties separating from off bypass on whether tricuspid stenosis was part of the problem after the cone repair and that is what led you to have to do the Glenn procedure? Can you comment on that, please?

Dr Liu. Yes, we have some cases. After the cone reconstruction procedure, if the RV pressure is high, then we also check it by using echocardiography. If there is some stenosis of the TV, we do the bidirectional Glenn procedure.

Dr del Nido. Because the alternative is simply to fenestrate the anterior leaflet, and this is what da Silva does. In fact, he makes large fenestrations in the anterior leaflet; he has not seen tricuspid stenosis, and we have not either. I am wondering whether this is a difference in the technique that you are applying versus what other centers are applying.

The final question I have for you, and the concern with routine use of a bicaval pulmonary shunt in these patients, is that many of these patients have a substantial right ventricle that can generate quite a bit of pulsatile flow. Have you seen problems with hyperpulsatility of the SVC? If you see that, what do you do about it?

Dr Liu. That is a good question. After the bidirectional Glenn procedure, we perform one and a half ventricle repair. It is very interesting. In our country we have a lot more of the RV problem than in western countries. Therefore we have a lot of RV problems, such as very severe pulmonary stenosis and tricuspid stenosis and hypoplastic right heart syndrome. Therefore we do some one and a half ventricle repairs.

In this group we did not find very severe pulsatility after SVC syndrome. We only had 4 or 5 patients temporarily this way. We

had another group in which found the SVC syndrome complication after one and a half ventricle repair. Sometime after the operation, maybe 3 or 4 months, we found the complication of SVC syndrome. We do the banding of the main pulmonary artery or between the main pulmonary artery and the bidirectional Glenn shunt. We put this banding in the right pulmonary artery.

Dr Florentino Vargas (*Buenos Aires, Argentina*). It was a very nice presentation, doctor.

I saw you included patients with Carpentier type D obstruction in whom you applied the procedure. If you see the anatomy in these patients, the atrialized portion is huge. The ventricularized part is really minimal. In most of those cases, the valves are adherent to the outflow tract. These are the cases in which the valve itself creates some degree of pulmonary obstruction. Therefore it is difficult for me to imagine, and I am really interested to know whether the procedure was applicable to these patients. Because these are

the patients in whom we are prone to consider in the part of the Ebstein anomaly that are susceptible to a univentricular type of path rather than this approach.

First, did this kind of procedure apply to the Carpentier-type subset of patients? Second, is there any patient with Ebstein anomaly in whom you consider that the anatomy was not suitable for the cone procedure?

Dr Liu. Even in patients with the classification of Carpentier type D, we also do the one and a half ventricle repair. I think maybe this is only the follow-up, and therefore maybe we need the long-term results.

What is the second question?

Dr Vargas. Are there any patients in whom you saw the anomaly, determined it was not the Ebstein anomaly, and you said: well, it is too bad. We go and do a procedure, just put him in the ventricular path, I don't think a repair can be done.

Dr Liu. In this series we did not find that.