

# De Vega Tricuspid Annuloplasty for Systemic Tricuspid Regurgitation in Children with Univentricular Physiology

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**Background and aim of the study:** Significant tricuspid valve regurgitation (TR) is problematic in children with univentricular physiology and a systemic tricuspid valve occasionally requiring tricuspid (systemic atrioventricular) valve replacement. Since 1998, the De Vega tricuspid annuloplasty technique has been applied for TR in these children.

**Methods:** Twelve children (median age 2.2 years; range: 6 months to 17 years) with moderate or severe systemic TR underwent a De Vega tricuspid annuloplasty during a bidirectional Glenn anastomosis (n = 3), Fontan procedure (n = 8) or aortic valve replacement late after a Fontan procedure (n = 1). Nine patients (75%) had prior Norwood palliation for hypoplastic left heart syndrome. Four patients had simultaneous repair of an abnormal tricuspid valve in addition to the De Vega procedure.

**Results:** There were no deaths during a mean follow up of  $2.0 \pm 1.4$  years (range: 6 months to 5.1 years).

The ultimate surgical 'repair' of children with univentricular physiology is a modified Fontan procedure. In children with a right ventricular-dominant circulation, the tricuspid valve functions as the systemic atrioventricular valve. Commonly, children who will eventually need a Fontan procedure will have significant tricuspid regurgitation (TR) when the tricuspid valve is the systemic atrioventricular valve (1). The presence of systemic TR in patients undergoing a Fontan procedure has classically been considered to be a relative risk factor, particularly in patients with hypoplastic left heart syndrome. The presence of mod-

erate or severe TR after a Norwood palliation has been shown to be a risk factor for mortality (2). One child required pacemaker implantation early after operation, and one child with a Glenn anastomosis underwent cardiac transplantation 21 months postoperatively. In the remaining 11 patients, the most recent echocardiogram showed mild or no TR in eight children, mild-to-moderate TR in one child, and moderate TR in two children. No child had symptomatic TR (including the two with moderate TR), significant tricuspid stenosis, or late pacemaker implantation.

**Conclusion:** The De Vega tricuspid annuloplasty safely provides excellent relief of systemic TR in children with univentricular physiology, with a majority of patients (73%) having mild or less residual TR at follow up examination. This simple technique is preferred to tricuspid (systemic) valve replacement in these children.

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Although it has been shown that children with significant systemic TR are able to undergo successful atrioventricular valve replacement (3), it would be preferable to avoid the use of a prosthetic valve in these growing children. Since 1998, the use of a simple De Vega annuloplasty technique in children with significant systemic TR has been explored in the hope of improving suitability for a Fontan procedure and avoiding valve replacement. The present report is a summary of the authors' experience in this respect.

## Clinical material and methods

### Patients

Between April 1998 and December 2002, 12 children with univentricular physiology with moderate or severe systemic TR underwent a De Vega tricuspid annuloplasty. TR was determined preoperatively with transthoracic echocardiography (TTE), or intraopera-

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tively with transesophageal echocardiography (TEE) using a semi-quantitative grading system as measured by spectral Doppler analysis; the study indicating the greater degree of regurgitation was used for classification. The TR was categorized as either absent, mild, moderate or severe (2). Mild TR was defined as a narrow regurgitant jet less than 1 mm which did not extend beyond an imaginary line one-third of the distance from the tricuspid valve annulus to the posterior wall of the right atrium. Moderate TR was defined as a regurgitant jet between 1 and 2 mm in diameter extending to the middle third of the right atrium. Severe TR extended broadly into the distal right atrium with a width greater than 3 mm.

The mean patient age at the time of operation was 2.2 years (range: six months to 17 years), and all children but one 17-year-old were aged three years or less. The patient characteristics are listed in Table I. Nine children had hypoplastic left heart syndrome, two had double-inlet left ventricle with ventricular septal defect and transposition of the great arteries, and one had congenitally corrected transposition of the great arteries with a hypoplastic double-outlet right ventricle and pulmonary stenosis. All children had undergone between one and three previous operations (mean  $2.0 \pm 0.6$ ). Nine children (75%) had a first stage Norwood procedure in infancy. Two of the other three children had systemic to pulmonary artery shunts as infants, and the third child had a balanced pulmonary circulation for which the first operation was a bidirectional Glenn anastomosis. Eight of the 12 children had their De Vega tricuspid annuloplasty at the time of a

modified fenestrated lateral tunnel Fontan procedure, and three children underwent a De Vega annuloplasty at the time of a bidirectional Glenn anastomosis. The final child (Patient 8 in Table I) was a 17-year-old who developed severe neo-aortic valve regurgitation as well as TR late after a Fontan procedure for hypoplastic left heart syndrome. He underwent a De Vega tricuspid annuloplasty at the time of his aortic valve replacement.

### Operative technique

The technique used for the De Vega tricuspid annuloplasty in children has been previously described for a tricuspid valve in the pulmonary ventricle (4). All operations were performed through a redo sternotomy on standard cardiopulmonary bypass (CPB) with bicaval cannulation. Hypothermic cardioplegic arrest was used in four patients, including the one who required aortic valve replacement and another who had patch enlargement of her residual coarctation following the first stage Norwood procedure at the time of a bidirectional Glenn anastomosis. In the remaining eight children the procedure was carried out during induced ventricular fibrillation.

The De Vega tricuspid annuloplasty was performed using a modification of De Vega's original technique (5). The tricuspid valve was inspected carefully for structural abnormalities. In four patients, the valve was structurally abnormal and required repair. This consisted of a combination of closure of abnormal clefts or shortening of elongated chordae. The tricuspid annulus was found to be dilated in all 12 patients.

Table I: Characteristics of the patient population.

Patient number	Diagnosis	Age at operation (years)	Number of prior operations	Other procedure at time of De Vega
1	TGA/DORV/PS	1.8	3	Fontan
2	HLHS	0.5	1	Glenn
3	HLHS	2.5	2	Fontan
4	DILV/VSD/Hypoplastic RV/TGA	2.5	2	Fontan; repair TV
5	HLHS	1.6	2	Fontan; repair TV
6	HLHS	2.0	2	Fontan
7	HLHS	2.6	3	Fontan
8	HLHS	17.0	2	AVR after Fontan
9	HLHS	0.5	2	Glenn; patch recurrent CoA
10	DILV/VSD/Hypoplastic RV/TGA	3.1	2	Fontan, repair TV
11	HLHS	3.3	2	Fontan
12	HLHS	0.6	1	Glenn; repair TV

AVR: Aortic valve replacement; CoA: Coarctation; DILV: Double-inlet left ventricle; DORV: Double-outlet right ventricle; HLHS: Hypoplastic left heart syndrome; PS: Pulmonary stenosis; RV: Right ventricle; TGA: Transposition of the great arteries; TV: Tricuspid valve.

After valve repair (if necessary), the tricuspid valve was tested for incompetence by injecting saline into the ventricle. A pledgetted annuloplasty suture of 3-0 or 4-0 polypropylene was started at the antero-septal commissure and sewn intermittently to the junction of the annulus and right ventricle along the anterior and posterior leaflets of the tricuspid valve until just past the postero-septal commissure. Another pledget was placed, and the suturing reversed along the posterior and anterior leaflets, 1-2 mm from the first row, and alternating the suture technique. That is, the suture bites on the second row were in where the suture was out on the first row, and vice versa. This continued until the original pledget at the antero-septal commissure was reached. The suture was tied down snugly over a Hegar dilator calibrated to 2-3 mm larger than the predicted appropriate pulmonary annulus size derived from published nomograms (6). The valve was again tested for competence by injecting saline into the ventricle. At this point, the remainder of the operation was completed. In eight patients, this consisted of a fenestrated lateral tunnel Fontan procedure, in three it was a bidirectional Glenn anastomosis, and in one patient it was an aortic valve replacement.

### Follow up

Tricuspid valve competence was evaluated in the operating room with TEE after repair, and before hospital discharge with TTE using the semi-quantitative grading system described above.

The children were followed with periodic cardiology evaluations and TTE as indicated clinically. The results of the most recent echocardiographic study were used for quantification of the degree of late tricuspid valve function.

### Results

There were no operative or late deaths at a mean follow up of  $2.0 \pm 1.4$  years (range: six months to 5.1 years). Patient 4, who had a double-inlet left ventricle, transposition of the great arteries and a restrictive ventricular septal defect with hypoplastic right ventricle, had undergone enlargement of his restrictive ventricular septal defect on two previous occasions. He developed heart block that required a permanent pacemaker. Patient 2, who had hypoplastic left heart syndrome and underwent a De Vega tricuspid annuloplasty at the time of her Glenn anastomosis, developed significant ventricular failure and underwent successful orthotopic cardiac transplantation 21 months postoperatively. An echocardiogram before transplantation showed the presence of moderate TR.

The echocardiography results for the remaining 11 patients are shown in Figure 1. On the early postoper-

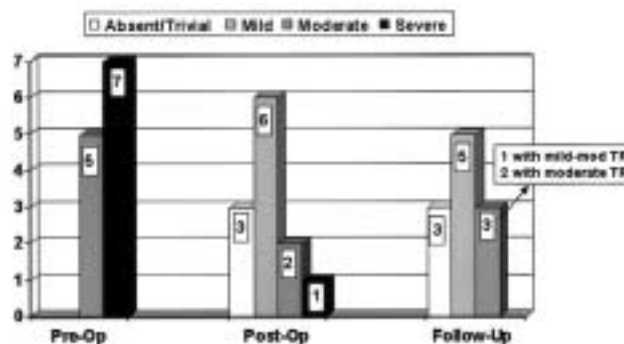


Figure 1: Semi-quantitative evaluation of tricuspid regurgitation (TR) by echocardiography preoperatively (Pre-Op), in the early postoperative period before hospital discharge (Post-Op), and at the latest follow up echocardiogram (Follow-Up). Preoperatively, all patients had moderate or severe TR, whereas 75% had mild or less regurgitation before hospital discharge. At the latest follow up, only three patients had more than mild regurgitation (one mild-to-moderate, two moderate). The one patient who underwent subsequent cardiac transplantation had moderate TR at the time of transplantation (not shown).

ative echocardiogram, two patients showed no improvement in the degree of TR (one with moderate, and one with severe TR). Three patients showed improvement of one grade in their TR; that is, from severe to moderate, or from moderate to mild. The remaining seven patients showed an improvement of more than one grade of TR. At early postoperative echocardiography, nine patients (75%) had mild or less regurgitation, one patient had mild to moderate regurgitation, one patient had moderate regurgitation, and the final patient had persistently severe regurgitation (Fig. 1).

The results obtained with the De Vega annuloplasty were relatively stable. The most recent follow up echocardiogram showed that in only one patient had the severity of regurgitation increased by a full grade (from mild to moderate); furthermore, the patient with postoperative severe regurgitation now had moderate TR. One of the patients with moderate regurgitation at early postoperative echocardiography now had mild regurgitation (Fig. 1). Neither of the two patients with moderate TR at follow up echocardiography was symptomatic.

At follow up, none of the patients demonstrated significant tricuspid stenosis, the need for late pacemaker implantation, or any need for further operation on the systemic tricuspid valve. Of the three children who underwent a De Vega tricuspid valve annuloplasty at the time of their bidirectional Glenn anastomosis, one underwent transplantation and the other two have been followed for less than one year and thus are

awaiting their Fontan procedure. Ventricular function at the latest follow up echocardiography was categorized as good or normal in 10 patients, slightly decreased in one patient, and poor in the one patient who required cardiac transplantation.

## Discussion

Systemic atrioventricular valve regurgitation in children undergoing the Fontan procedure is a common occurrence. Regurgitation was found in 41% of 242 patients undergoing a Fontan procedure in a report from Imai et al. (1) Approximately half of these patients (n = 49) had either moderate or severe TR. Even when an attempt was made at repair using a circular annuloplasty technique, the patients with significant systemic atrioventricular valve regurgitation had a higher early mortality with the Fontan procedure compared with those who did not.

When the systemic atrioventricular valve is a tricuspid valve, regurgitation is particularly problematic. Barber and colleagues found moderate or severe TR in 15% of children investigated after a first stage Norwood procedure for palliation of hypoplastic left heart syndrome (2). Tricuspid regurgitation in these patients was found to be a risk factor for death, and these authors recommended either an early annuloplasty or tricuspid valve replacement if significant TR occurred. The Philadelphia group found moderate or severe TR in five of 59 children after the first stage Norwood procedure. This was shown to be a significant risk factor for death at the time of the Fontan procedure. All three patients with severe TR died, and one of the two patients with moderate TR died at the time of the Fontan procedure (7).

It seems, therefore, that it is important to address significant systemic tricuspid valve regurgitation in children with univentricular hearts who eventually will require a Fontan procedure. Tricuspid regurgitation is a significant problem in children with congenitally corrected transposition who, although having the potential for a biventricular repair, have the tricuspid valve as the systemic atrioventricular valve. Certainly, tricuspid valve replacement can be carried out successfully in these children, with good results (3). Attempts at valve repair sometimes are discouraging, however. Acar and colleagues in Paris found that repair of a systemic tricuspid valve in congenitally corrected transposition was uniformly unsuccessful in their hands (8).

Despite these discouraging results with congenitally corrected transposition, the present authors still felt it preferable to attempt valve repair rather than valve replacement in children with univentricular physiology and systemic TR. The Michigan group has published data previously on this problem (9,10). In

addition to the repair of abnormal valve leaflets when necessary, a partial annuloplasty technique was used which essentially involved obliterating the posterior leaflet of the tricuspid valve. The most recent report of these authors reviewed 28 children undergoing repair of a systemic tricuspid valve among 475 patients after a first stage palliation for hypoplastic left heart syndrome (11). Of these 28 children, half had a tricuspid valve repair at the time of a hemi-Fontan procedure, and the other half had their repair at the time of the Fontan procedure. Ohye et al. (11) determined that children who had a good initial outcome had an excellent survival (94%), whereas those with an unsatisfactory repair in the early postoperative period had a very poor survival (20%). Furthermore, the early postoperative repair correlated closely with the long-term follow up of valve function. If the child had a successful repair, then preoperative TR did not appear to be a risk factor for death (9,11).

Previously, the present authors have reported the application of the De Vega tricuspid annuloplasty to children with TR associated with the pulmonary ventricle (4). As many children with systemic TR and univentricular physiology basically have a dilated valve annulus, it was reasoned that application of this simple technique to children with systemic tricuspid valve regurgitation would be useful. In the present series, the De Vega annuloplasty was efficacious, resulting in nine out of the 12 patients (75%) having mild or less TR at the time of hospital discharge. There were no deaths in this group, nor was there any significant tricuspid stenosis (Fig. 1). One patient did require a pacemaker early after operation, but this child (Patient 4, Table I) with double-inlet left ventricle and restrictive ventricular septal defect had previous enlargement of the septal defect on two separate occasions. It is unclear if his postoperative heart block was related to the De Vega annuloplasty, or to the previous ventricular septal defect enlargements.

Based on an average follow up of 1.5 years, the De Vega tricuspid annuloplasty seems to be durable. Only one patient (Patient 2, who required cardiac transplantation) had an increase of at least one grade of TR on the echocardiogram obtained during the postoperative period compared with the echocardiogram obtained at follow up. In fact, two patients had a full grade decrease in regurgitation at follow up, including one who had severe regurgitation in the immediate postoperative period and one who had moderate regurgitation in the postoperative period.

Naturally, only time will tell if this beneficial effect is longstanding. Of the three children who had their De Vega annuloplasty at the time of the Glenn procedure, one underwent transplantation and the other two have been followed for less than one year and thus are

awaiting their Fontan procedure. Certainly, if this technique is durable, then it is far more preferable to systemic atrioventricular valve replacement with the attendant need for re-replacement for biological valves or anticoagulation risks with mechanical valves.

The particular technique of De Vega tricuspid annuloplasty seems to be well suited in this set of patients. The circular annuloplasty technique described by Imai et al. appeared to be associated with an increased early operative mortality, although it is certainly not clear whether the specific technique was the risk factor or if it was simply the presence of preoperative atrioventricular valve regurgitation (1). Ohye and colleagues (11) described the partial annuloplasty technique with obliteration of the posterior leaflet, and 63% of their patients had an early favorable result compared with 75% among the present patients. Unfortunately, the patient numbers in the present series were too small for this difference to be significant, but the findings showed great promise.

Although the present authors feel that the cause of regurgitation in their patients was usually due to right ventricular distension with consequent annular dilatation, a certain percentage of the patients did have abnormal valves. Four of the 12 patients required tricuspid valve repair, which compared well with five of eight patients who also required tricuspid valve repair in the series by Reyes et al. (10). Interestingly, all four patients in the present series with tricuspid valve repair in addition to a De Vega annuloplasty had mild or less TR at the latest follow up examination.

*In conclusion*, the De Vega tricuspid annuloplasty appears to be an efficacious and relatively simple technique for reducing moderate to severe systemic tricuspid valve regurgitation in children with univentricular physiology undergoing a Glenn or Fontan procedure. The early results of the present series were favorable, with no mortality, only one pacemaker required, and no early or late tricuspid stenosis. At follow up, the repair appeared to be reasonably durable. The benefits of avoiding systemic atrioventricular valve replacement are clear.

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