Interactive CardioVascular and Thoracic Surgery

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*Interact CardioVasc Thorac Surg* 2010;10:900-905; originally published online Mar 5, 2010;
DOI: 10.1510/icvts.2009.223982

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INTERACTIVE CARDIOVASCULAR AND THORACIC SURGERY

Interactive CardioVascular and Thoracic Surgery 10 (2010) 900-905

www.icvts.org

Institutional report - Congenital

# The impact of the length between the top of the interventricular septum and the aortic valve on the indications for a biventricular repair in patients with a transposition of the great arteries or a double outlet right ventricle☆

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Received 1 October 2009; received in revised form 3 February 2010; accepted 4 February 2010

**Abstract**

The purpose of this study was to establish a useful cut-off level for performing an original Rastelli-type operation in patients with transposition of the great arteries (TGA)/ventricular septal defect (VSD) or double outlet right ventricle (DORV). A total of 43 patients with TGA/VSD or DORV who underwent an original Rastelli-type operation in this institute between March 1993 and January 2009 were reviewed retrospectively. These patients were divided into two groups using the length between the top of the interventricular septum and the aortic valve (IVS–AV length); Group A; IVS–AV length <80% of normal left ventricular end-diastolic diameter (LVDd). Group B; IVS–AV length ≥80% of normal LVDd. Group A had a significantly better survival than Group B (100% vs. 56%, $P=0.001$). The cardiac event-free survival were 89.1% at 7.2 years in Group A and 26.3% at 8.4 years in Group B ($P<0.0001$). The Group B had a higher incidence of left ventricular outflow tract obstruction (LVOTO; 3% vs. 33%, $P=0.02$). The IVS–AV length was found to be a significant risk factor for mortality and LVOTO. The IVS–AV length should, therefore, be taken into consideration when selecting the optimal surgical procedures for these patients.


*Keywords:* Transposition of great vessels; Double outlet right ventricle; Left ventricular outflow obstruction; Cardiac surgical procedures; Ventricular septal defect

## 1. Introduction

Patients who have a transposition of the great arteries (TGA) + ventricular septal defect (VSD) with pulmonary obstruction or double outlet right ventricle (DORV) with pulmonary obstruction are usually treated with an original Rastelli-type operation that includes intraventricular rerouting to creating a left ventricular outflow tract (LVOT) and reconstruction of the right ventricular outflow tract (RVOT) [1]. Patients undergoing an original Rastelli-type operation often require a long intraventricular rerouting patch, and it may be a risk factor for death, left ventricular outflow tract obstruction (LVOTO), and heart failure [2]. However, no study has yet clearly established the indications for an original Rastelli-type operation or identified which patients are candidates for a more complicated biventricular repair, such as the Réparation à l'Etage Ventriculaire (REV) operation [3], the Nikaidoh procedure [4] and a half-turned truncal switch operation [5], as well as which patients are candidates for the Fontan operation. One study reported the mid-term results of the Nikaidoh procedure to be superior to the Rastelli operation [6] although the Nikaidoh procedure had a high early mortality [6, 7]. In addition, the long-term outcome of the Fontan operation may be better than a complex biventricular repair [8, 9]. Therefore, elucidating the limitations of the original Rastelli-type operation is a very important issue. The purpose of this study was to describe the outcomes in a patient who underwent an original Rastelli-type operation and to verify whether the length of the intraventricular rerouting patch really affects the outcomes.

## 2. Materials and methods

### 2.1. Patients

This study was approved by the Institutional Research Ethics Board at the Okayama University Hospital (OUH), and patient consent was waived. The records of patients who underwent an original Rastelli-type operation in OUH between June 1992 and April 2008 were reviewed retro-

☆ Presented at the 23rd Annual Meeting of the European Association for Cardio-thoracic Surgery, Vienna, Austria, October 18–21, 2009.

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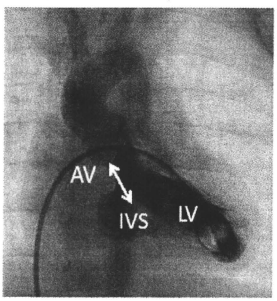
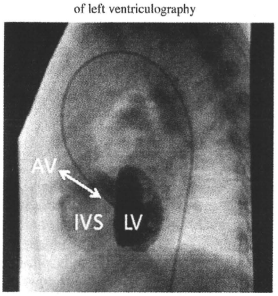
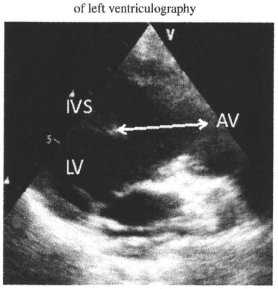
spectively. Patients without a sufficient length between the tricuspid valve and the pulmonary valve, patients with DORV with subpulmonary VSD, patients with inlet VSD, and patients with abnormal tricuspid chordae which cross the area for LVOT were excluded in this study.




Frontal view
of left ventriculography




Lateral view
of left ventriculography




Parasternal long axis view
of cardioechography

Fig. 1. The measurement of the length between the top of the interventricular septum and the aortic valve (IVS–AV length). The length between the top of the interventricular septum and the aortic valve (IVS–AV length) was measured by the parasternal long axis view of echocardiography and by either a frontal view or a lateral view of a catheter examination. AV, aortic valve; IVS, interventricular septum; LV, left ventricle.

### 2.2. Measurement of the IVS–AV length

The length between the top of the interventricular septum and the aortic valve (IVS–AV length) was regarded as the length of the intraventricular rerouting and was measured by the parasternal long axis view of echocardiography and by either the frontal view or lateral view of catheter examination (Fig. 1). The maximum lengths of these measured values were adopted for this study.

### 2.3. Surgical procedures

All of the operations were performed using a conventional continuous flow cardiopulmonary bypass and mild to moderate hypothermia. Cardiac arrest was obtained with cold (4 °C) crystalloid cardioplegia after aortic cross-clamping. A right atriotomy was performed. Reconstruction of the LVOT was performed with a polytetrafluoroethylene (PTFE) or Dacron sauvage patch. A pulmonary arteriotomy was performed in patients with pulmonary stenosis (PS). A right ventriculotomy was performed in patients with pulmonary atresia (PA). The muscles protruding into the RVOT were resected in patients with PS. Atrial rerouting was performed after VSD closure in patients with an atrioventricular discordance. The method of RVOT reconstruction depended on the anatomies of each patient. When patients had a PS that did not require a transannular patch, a pulmonary valvotomy was performed, then, if necessary, the pulmonary artery was enlarged with a fresh autologous pericardial patch. When patient had a PS which required a transannular patch, the incision of pulmonary arteriotomy was extended to the RVOT. Thereafter, the RVOT was reconstructed using a fresh autologous pericardial patch with a PTFE monocuspid valve. When patients had PA, the site of the right ventriculotomy was determined according to the intracardiac anatomy of each patient. An artificial conduit was interposed between the right ventricle (RV) and the central pulmonary artery.

### 2.4. Statistical analysis

Binary data or categorical data (except for patients' diagnosis) were analyzed using Fisher's exact test. Pearson's $\chi^2$-test was used for comparison of patient's diagnosis. Student's *t*-test was used for comparison of continuous data. Tricuspid regurgitation (TR) and pulmonary regurgitation (PR) were compared with the Mann–Whitney *U*-test. The correlation between the IVS–AV length and aortic cross-clamp time (ACCT) was evaluated using a bivariate linear regression analysis. Data are expressed as the mean $\pm$ standard deviation (S.D.) or median and range as appropriate. A $P<0.05$ was considered to be significant. The normal left ventricular end-diastolic diameter (LVDd) and normal aortic valve (AV) diameter means the average LVDd and the average AV diameter in normal population that were described in a previously published test book [10], respectively. The definition of early death is death which occurs within 30 days after the operation.

## 3. Results

### 3.1. Patients' diagnosis and demographics

Forty-three patients were entered in this study. The main cardiac diagnoses were DORV in 26, TGA+VSD in five, and congenitally corrected transposition of the great arteries (ccTGA)+VSD in 12. A diagnosis of DORV was made with application of the 50% rule [11]. Thirteen of 26 patients with DORV had tetralogy of Fallot-type DORV. All of the other patients with DORV except for one patient with a doubly committed VSD had a subaortic VSD. All of those patients had either PA (including one patient with non-confluent pulmonary artery) or PS. The median age at the Rastelli-type operation was 28 months (range, 0–430). The median body weight was 11.3 kg (range, 2.2–60.0).

### 3.2. Surgical data

All of the ccTGA patients underwent a Rastelli-type double-switch-operation with 11 Senning and one Mustard operation. The RVOT was reconstructed with an artificial valve conduit in 18, a homograft in two, transannular patch plasty with a PTFE monocuspid valve in seven, simple patch plasty of the main pulmonary artery in three, pulmonary valvotomy only in 12, and direct anastomosis between the main pulmonary artery and the RV in one. The associated procedures were VSD enlargement in 12 (conal septal resection in eight, anterosuperior VSD enlargement in three, conal septal resection and inferior VSD enlargement in one), pulmonary arterial plasty in 10, tricuspid valve repair in three (a commissuroplasty in one, division of an abnormal tricuspid chorda in one, suspension of the tricuspid annulus in one with mild Ebstein), and transfer of the left pulmonary artery in one.

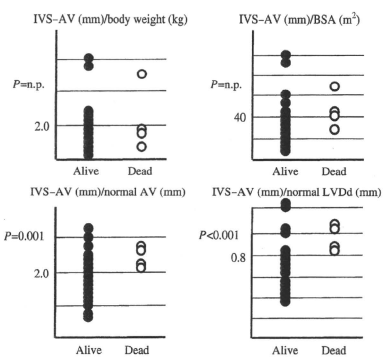



Fig. 2. Several ratios for expressing the relative length of IVS–AV. Several references were consulted to create a ratio for expressing the relative length of IVS–AV against the body growth; the body weight (IVS–AV length/patient's body weight), the body surface area (IVS–AV length/patient's body surface area), normal LVDd value (IVS–AV length/normal LVDd), and normal aortic valve diameter (IVS–AV/normal aortic valve diameter). The value of the vertical axis shows the cut-off points for analysis. The IVS–AV length/normal LVDd ratio with a cut-off point of 0.8 (80% of normal LVDd) appeared to have the most significant influence on mortality. AV, aortic valve diameter; BSA, body surface area; IVS–AV length, the length between the top of the interventricular septum and the aortic valve; LVDd, left ventricular end-diastolic diameter.

Table 1
Risk analysis regarding death

| | Alive ($n=39$) | Dead ($n=4$) | $P$-value |
|---|---|---|---|
| Age <12 months | 5 | 1 | n.s. |
| IVS–AV length ≥80% of normal LVDd | 0 | 4 | <0.001 |
| Postoperative LVOTO | 2 | 2 | 0.004 |
| CPBT ≥300 min | 1 | 3 | <0.001 |
| ACCT ≥180 min | 1 | 3 | <0.001 |
| TR≥III | 0 | 1 | n.s. |
| RVEDV ≥120% of $n$ | 3 | 1 | n.s. |
| LVEF <60% | 8 | 1 | n.s. |
| RVEF <50% | 4 | 0 | n.s. |
| Female gender | 11 | 3 | n.s. |
| Diagnosis | TGA; 5, cTGA; 9, DORV; 25 | cTGA; 3 DORV; 1 | n.s. |
| VSD enlargement | 11 | 1 | n.s. |

IVS–AV length, the length between the top of the interventricular septum and aortic valve; LVDd, left ventricular end-diastolic diameter; LVOTO, left ventricular outflow tract obstruction; CPBT, cardiopulmonary bypass time; ACCT, aortic cross-clamp time; TR, tricuspid regurgitation; RVEDV, right ventricular end-diastolic volume; LVEF, left ventricular ejection fraction; RVEF, right ventricular ejection fraction; TGA, transposition of the great arteries; cTGA, corrected transposition of the great arteries; DORV, double outlet right ventricle; VSD, ventricular septal defect.

### 3.3. Postoperative outcomes

There was one early death and three late deaths. The early death was due to low output syndrome (LOS). The late deaths were caused by LOS in one, ventricular fibrillation in one, and heart failure with a subaortic stenosis in one. Two of those three patients were hospital deaths (LOS and ventricular fibrillation). The mean follow-up period for the 42 early survivor was 69±54 months (1–201). Four patients experienced LVOTO, which was defined with ≥2.0 m/s blood flow in the LVOT by echocardiography or ≥5 mmHg pressure gradient by catheter examination. Late postoperative tachyarrhythmias occurred in three patients; supraventricular tachyarrhythmia in one, ventricular fibrillation in one, and ventricular tachycardia in one. Eight patients underwent surgical re-intervention; re-do RVOT reconstruction in five, release of a pulmonary venous obstruction (PVO) in one, closure of an orifice of the LVOT aneurysm in one, and closure of a residual VSD in one.

### 3.4. Risk analysis for mortality

The IVS–AV length was expressed with a ratio which was calculated from the measured maximum IVS–AV length (mm) divided by the normal LVDd (mm). Several references were consulted to create the ratio for expressing the relative length of IVS–AV against the body growth. Fig. 2 shows the results. The IVS–AV length/normal LVDd ratio with cut-off point of 0.8 (80% of normal LVDd) appeared to have the most significant influence on mortality. Table 1 shows the results of a risk analysis for mortality.

Table 2
Comparison of the preoperative and surgical data between Group A and Group B

| | Group A ($n=34$) | Group B ($n=9$) | $P$-value |
|---|---|---|---|
| Age (months) | Median 28 (5–430) | Median 29 (0–353) | n.s. |
| IVS–AV length (mm) | 16.6±4.7 (8.0–24.5) | 25.9±10.1 (10.9–41.9) | 0.037 |
| IVS–AV length (% of normal LVDd) | 56±12 (40–79) | 101±15 (84–124) | <0.001 |
| Estimated LVOT diameter (% of normal AV) | 106±56 (56–168) | 126±47 (46–177) | n.s. |
| Female | 11 | 3 | n.s. |
| Diagnosis | TGA; 4, cTGA; 8, DORV; 22 | TGA; 1, cTGA; 4, DORV; 4 | n.s. |
| Body weight (kg) | Median 10.6 (3.9–60.0) | Median 13.3 (2.2–44.5) | n.s. |
| Height (cm) | Median 81 (54–168) | Median 96 (45–152) | n.s. |
| BSA ($m^2$) | 0.60±0.34 (0.23–1.63) | 0.57±0.39 (0.16–1.38) | n.s. |
| CTR (%) | 55.6±5.1 (46–67) | 58.6±3.5 (54–63) | n.s. |
| LVEF (%) | 66±9 (44–80) | 59±8 (44–75) | n.s. |
| RVEF (%) | 65±8 (43–81) | 58±10 (44–77) | n.s. |
| TR | No; 1, Tirv; 20, Mild; 13 | Triv; 4, Mild; 4, Sev; 1 | n.s. |
| RVEDV (%) | 124±38 (67–214) | 138±36 (83–191) | n.s. |
| CPBT (min) | Median 152 (76–398) | Median 148 (88–381) | n.s. |
| CPBT ≥300 min | 1 | 3 | 0.02 |
| ACCT (min) | Median 107 (55–260) | Median 116 (38–186) | n.s. |
| ACCT ≥180 min | 2 | 4 | 0.01 |
| VSD enlargement | 9/34 | 3/9 | n.s. |

IVS–AV length, the length between the top of the interventricular septum and aortic valve; LVDd, left ventricular end-diastolic diameter; LVOT, left ventricular outflow tract; AV, aortic valve; TGA, transposition of the great arteries; cTGA, corrected transposition of the great arteries; DORV, double outlet right ventricle; BSA, body surface area; CTR, cardio-thoracic ratio; LVEF, left ventricular ejection fraction; RVEF, right ventricular ejection fraction; TR, tricuspid regurgitation; RVEDV, right ventricular end-diastolic volume; CPBT, cardiopulmonary bypass time; ACCT, aortic cross-clamp time; VSD, ventricular septal defect.

Table 3
Comparison of the postoperative outcomes between Group A and Group B

| | Group A ($n=34$) | Group B ($n=9$) | $P$-value |
|---|---|---|---|
| Death | 0/34 | 4/9 | 0.001 |
| Cardiogenic event (except for re-do RVOTR and PVO) | 2/34 | 6/9 | <0.001 |
| Postoperative LVOTO | 1/34 | 3/9 | 0.02 |
| Postoperative TR | Triv; 25, Mild; 9 | Triv; 7, Mild; 1, Mod; 1 | n.s. |
| Postoperative PR | No; 3, Triv; 16, Mild; 7, Mod; 8 | No; 0, Triv; 8 | n.s. |

RVOTR, right ventricular outflow tract reconstruction; PVO, pulmonary venous obstruction; LVOTO, left ventricular outflow tract obstruction; TR, tricuspid regurgitation; PR, pulmonary regurgitation.

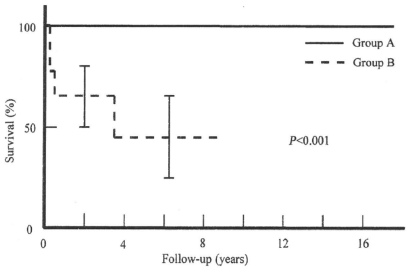



Fig. 3. Estimated survival of Group A and Group B. The estimated survival was significantly lower in Group B than in Group A (100% at 16.8 years in Group A vs. 44.4% at 8.4 years in Group B, $P<0.001$).

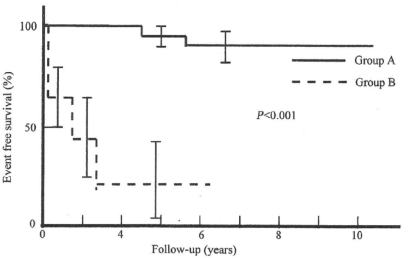



Fig. 4. Estimated cardiac event-free survival (other than a re-do RVOT reconstruction and a pulmonary venous obstruction) of Group A and Group B. The cardiogenic event-free survival (other than a re-do RVOT reconstruction and a pulmonary venous obstruction) was also significantly lower in Group B than in Group A (89.7% at 16.8 years in Group A and 26.3% at 8.4 years in Group B ($P<0.001$). RVOT, right ventricular outflow tract.

The longer IVS–AV length tended to have positive correlation with longer ACCT (R=0.763 and $P=0.132$ in TGA patients; R=0.529 and $P=0.09$ in ccTGA patients; R=0.303 and $P=0.160$ in DORV patients).

### 3.5. Group A vs. Group B

All of these patients were divided into two groups according to the IVS–AV length; Group A; IVS–AV length of <80% of normal LVDd. Group B; IVS–AV length of ≥80% of normal LVDd. Table 2 shows the results of comparisons of the preoperative, operative, and postoperative data between Group A and Group B. Table 3 describes the comparison of

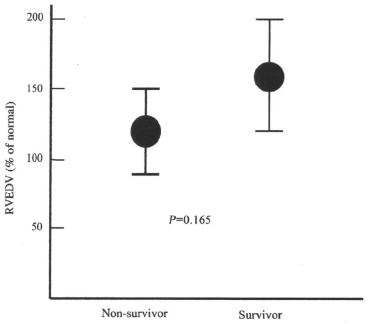



Fig. 5. Influence of the preoperative right ventricular end-diastolic volume in patients of Group B. The survivors in Group B tended to have a larger right ventricular end-diastolic volume than non-survivors (156±29% vs. 119±35%, $P=0.165$). RVEDV, right ventricular end-diastolic volume.

the postoperative outcome between Group A and Group B. The cardiac events mean left heart failure either with or without LVOTO, re-operations related with the LVOT, and arrhythmias. Re-do RVOT reconstructions and a reoperation due to PVO were excluded because these reoperations were thought to not be related with the IVS–AV length. Fig. 3 shows that the estimated survival was significantly lower in Group B than in Group A. Fig. 4 shows that the cardiogenic event-free survival (except for re-do RVOT reconstructions and a PVO) was also significantly lower in Group B than in Group A. In addition, the survivors in Group B tended to have a larger right ventricular end-diastolic volume (RVEDV) than the non-survivors (Fig. 5).

## 4. Discussion

Lacour-Gayet et al. previously noted that long left ventricular rerouting tunnel may be a risk factor for death, LVOTO, and heart failure in patients who underwent a Rastelli operation [2]. However, there is no evidence to support this hypothesis. This study showed that a long IVS–AV length was an important risk factor for mortality, cardiogenic postoperative events, and LVOTO in patients who underwent the original Rastelli-type operation. The results of this study will provide justification for the selection of a repair type in this patient population.

This study used a ratio of the IVS–AV length to the normal LVDd to express the relative length of the IVS–AV against the body growth. However, the ratio of IVS–AV length to the normal AV diameter with cut-off point of 200% of normal AV diameter was also found to be significant difference on the mortality. However, using the LVDd seemed to be more sensitive than using the normal AV diameter. There may be a discrepancy between the growth of the LVDd and the growth of the AV diameter.

This study found that cardiopulmonary bypass time (CPBT) of ≥300 min, ACCT of ≥180 min, and postoperative LVOTO were also risk factors for mortality, however, the IVS–AV length seemed to have a large influence on these factors. In addition, the IVS–AV length seemed to have a positive correlation with ACCT. These results indicated that there was a strong relationship between the IVS–AV length and these intraoperative/postoperative factors. Therefore, the IVS–AV length is the causative preoperative factor for mortality.

The patients in the current series with an RVEDV of ≥120% of the normal RVEDV in Group B tended to have better survival than patients with that of <120% of normal RVEDV in Group B. A large RV may contribute to reduce the risk of long rerouting patch with reducing the risk of postoperative LVOTO because a large RV allows the placement of a sufficiently larger rerouting patch in the RV without impairing the function of the RV.

There are some limitations in this study. First, this study was conducted retrospectively. Therefore, not all of the IVS–AV lengths measurements were performed both with catheter and echocardiography. Second, an appropriate multivariate analysis could not be performed due to the small number of the patients. Third, this study included patients with a very diverse anatomical spectrum of the heart. With these limitations in this study, the influence of the IVS–AV length for an original Rastelli-type operation should be re-evaluated with a multivariate analysis in patient populations with each respective diagnosis in the future.

In conclusion, the IVS–AV length was found to be a significant risk factor for mortality and postoperative cardiogenic events. However, a larger RVEDV may somewhat compensate for the risk of mortality. The IVS–AV length should be taken into consideration when selecting the optimal surgical procedures for these patients.

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## Conference discussion

***Dr. B. Brawn*** *(Birmingham, UK)*: Chairmen, in typical fashion, Shunji Sano's group has focused on a pertinent point to try and indicate whether biventricular repair will reduce the problems with left ventricular outflow tract in this group of patients.

It seems that the longer the length of the top between the ventricular septum and the aortic valve as indicated in your diagram, the worse the outcome.

This is retrospective that you saw on echocardiograms. There is a mixed bag of diagnoses from double outlet to ccTGAs.

It would be fantastic if this worked, and as you pointed out, it's important to do a prospective study. These were all postoperative results. I congratulate the author on their innovative approach to this group of patients.

I have a few comments or a question I should say. Are you saying that the length of the left ventricular outflow tract that you measure from the top of the septum to the aortic valve perhaps is also related to postoperative left ventricular outflow tract obstruction? Was there left ventricular outflow tract obstruction in that group more than in the ones with the shorter patch?

***Dr. Fujii:*** Outflow tract obstruction? Your question is how to measure the outflow tract obstruction?

***Dr. Brawn:*** No. Was there left ventricular outflow tract obstruction postoperatively?

***Dr. Fujii:*** Yes.

***Dr. Brawn:*** Okay.

***Dr. S. Sano*** *(Okayama, Japan)*: I do answer all your questions.

If the length between the IVS and the aorta is longer, then the incidence of left ventricular outflow tract obstruction in the long-term is much higher.

***Dr. Brawn:*** And could you comment, please, on the benefit, if any, of including ccTGAs in this group of patients, and in particular, the relationship between the proportion of problems between those that have pulmonary stenosis and those that have pulmonary atresia. Because I think Viktor pointed out earlier when you have pulmonary atresia, the VSD is often more better aligned to the aorta.

***Dr. Sano:*** First, we try to analyze all the patients who require a longer intraventricular tunnel patch, so that's why we include the corrected TGA.

We know that the pulmonary stenosis and pulmonary atresia patients are a little bit different. There's also a difference between the Asian population and also the applications.

In the Asian population, the VSDs, to me, my experience, it's much bigger. And, also, some of the patients are very much remote.

***Dr. V. Hraska*** *(Sankt Augustin, Germany)*: I am wondering what is the clinical impact of your conclusions? Would you consider a different type of surgery in order to avoid the very long intraventricular tunnel? I am asking because I'm not sure if you can simplify this problem only to the lengths of the patch or to the length of the intraventricular tunnel, which of course plays, as you demonstrated, an important role.

The additional point is the shape of the tunnel itself which can be extremely important in the long-term. Can you comment on that?

***Dr. Sano:*** From our analysis, as you know, if the intraventricular rerouting patch is long, then non-functioning left ventricular cavities increase compare to the functioning left ventricular cavity. That's why the long-term result is not as good as the other group, I imagine. We try to analyze more details about these things, but there is no good evaluation method.

And also, we try to put a little bit bigger patch to avoid left ventricular outflow tract obstruction. That requires a larger right ventricular end-diastolic volume. But I think the long-term result may be the same because we have non-functioning cavities. If the patient has a long patch, we now change our policy. We do a Fontan strategy rather than doing a double switch because the Fontan result is quite good.

### eComment: Parameters of the channel between the left ventricle and the aortic valve in Rastelli-type operation

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**doi:**10.1510/icvts.2009.223982A

The authors touched upon an important question concerning the parameters of the heart and its structures during the operation for biventricular correction of complex congenital heart defects [1].

It is generally known that the Rastelli-type operation includes the creation of a channel within the right ventricular cavity serving for the connection of the left ventricle with the aortic valve. The subject of this study was the length of the channel formed between the left ventricle and the aorta (the length between the top of the interventricular septum and the aortic valve) and the influence of this parameter on the mortality and the frequency of postoperative left ventricular outflow tract obstruction (LVOTO). The authors concluded that the length of the channel between the left ventricle and the aorta is a reliable factor of mortality and LVOTO.

While being confident upon the veracity of the obtained results, we would like to discuss one problem. It concerns the spectrum of the anomalies included in the study. For a good reason, the authors have excluded patients with DORV with inlet ventricular septal defect (VSD), as well as patients with abnormal tricuspid chordae which cross the left ventricular outflow tract, as the correction of these DORV variants is more complicated and the formation of a direct channel between the left ventricle and the aorta in such cases as a rule is impossible [2, 3].

However, the material of the study included patients with congenitally corrected transposition of the great arteries (ccTGA) + VSD who underwent more complicated surgical repair (double-switch-operation) in comparison with the Rastelli-type operation for DORV with subaortic VSD. From our point of view, these groups are different in complexity and volume of surgical repair. As the paper states, three out of four dead patients had corrected ccTGA [3/12 (25%)], and the fourth one had DORV [1/26 (3.8%)]. Hence, the mortality in these groups is also different. It would be interesting to know the interventricular septum and the aortic valve length depending on the nosology (ccTGA, DORV), and to correlate it with the results of correction after the calculation of Z-score value from normal left ventricular end-diastolic diameter.

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**The impact of the length between the top of the interventricular septum and the aortic valve on the indications for a biventricular repair in patients with a transposition of the great arteries or a double outlet right ventricle**

Yasuhiro Fujii, Yasuhiro Kotani, Masami Takagaki, Sadahiko Arai, Shingo Kasahara, Shin-ichi Otsuki and Shunji Sano

*Interact CardioVasc Thorac Surg* 2010;10:900-905; originally published online Mar 5, 2010;
DOI: 10.1510/icvts.2009.223982

## DISCUSSION

Intracardiac lipomas usually present with encapsulation and have no potency of infiltrating adjoining tissues. In this case, the tumor had no encapsulation and theoretically was able to infiltrate the myocardium, which it did.

Lipomas are very slow-growing tumors, and this case also shows the enormous adaptive capacity of a normal heart as the patient had no cardiac symptoms except the premature ventricular contractions with a huge tumor in the left ventricle.

We believe this is the first report of a benign cardiac tumor behaving as a malignant tumor infiltrating myocardium.

All valvular tumors should be removed regardless of size or symptoms.[1] In this case, the surgery was curative as the tumor was completely resected and no recurrences have been seen.

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# Fontan completion in patient with pulmonary artery sling associated with hypoplastic left heart syndrome

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Pulmonary artery (PA) sling is an uncommon congenital anomaly in which the left PA arises from the posterior aspect of the right PA, forming a sling around the trachea.[1] Current surgical approach (ie, reimplantation of the left PA via median sternotomy under cardiopulmonary bypass [CPB]), offers favorable early and late outcomes.[2,3] Although approximately one-third of patients with PA sling have associated cardiac anomalies, only 1 patient reported in late 1950s had single-ventricle anomaly[1]; therefore, surgical implications of this entity in patients with functional univentricular heart have never been discussed. We herein describe a patient with asymptomatic PA sling associated with hypoplastic left heart syndrome (HLHS) who had staged Fontan completion.

## CLINICAL SUMMARY

A 2-day-old girl with the body weight of 1.9 kg was diagnosed with HLHS with persistent left superior vena cava. The existence of PA sling was suspected on preoperative echocardiography. No respiratory symptoms were noted. She had modified Norwood procedure using a right ventricle–to–pulmonary artery (RV-PA) shunt at 3 days of age. The left PA originating from the right PA was detected. The main PA was divided and was used for aortic arch reconstruction. The distal side of the RV-PA shunt was anastomosed to the right PA with a noncuffed 5-mm polytetrafluoroethylene graft (Gore-Tex expanded polytetrafluoroethylene graft, W. L. Gore & Associates, Inc, Flagstaff, Ariz). The postoperative course was uneventful. Cardiac catheterization at 4 months of age revealed mean PA pressure of 13 mm Hg in the right PA and 9 mm Hg in the left PA. The right PA was 6 mm in diameter and the left was 3.1 mm in diameter with hypovascularity in its lung field (Figure 1, *A*). The Nakata index was 163 $mm^2/m^2$. PA resistance was 2.75 W/U. Bidirectional Glenn (BDG) anastomosis on the right side was performed at 6 months of age through a median sternotomy under CPB. Arterial desaturation persisted after surgery, and we additionally performed BDG on the left through a left thoracotomy, resulting in increase in $SpO_2$. The RV-PA shunt was left open. Fontan operation was performed at the age of 2 years and 2 months, at an earlier timing than expected, due to a restrictive interatrial communication. The right PA was 7.2 mm in diameter and the left was 6.3 mm in diameter. The Nakata index was 218 $mm^2/m^2$. The left PA was untreated and the patient has had no respiratory symptoms throughout the staged

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Disclosures: None.
Received for publication May 8, 2009; accepted for publication July 5, 2009; available ahead of print Aug 26, 2009.
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J Thorac Cardiovasc Surg 2010;140:e12-3
0022-5223/$36.00

doi:10.1016/j.jtcvs.2009.07.021

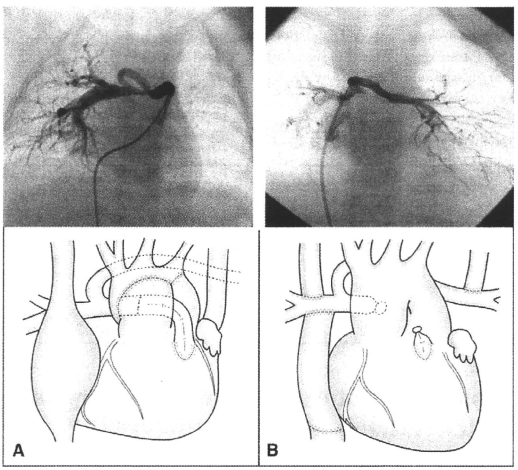


**FIGURE 1.** A, Pulmonary artery *(PA)* angiogram before bidirectional Glenn anastomosis showing that the left PA originated from the right PA and formed the sling around the trachea. No stenosis but hypovascularity was observed in the left pulmonary artery. B, Angiogram 2 years after Fontan operation showing reasonable left PA growth.

palliation. No stenosis was found in the left PA on angiography (Figure 1, *B*), and the nuclear pulmonary perfusion scan showed that the right-to-left perfusion ratio was 2.3 to 1 at the follow-up period of 4 years.

## DISCUSSION

This is the first reported patient with PA sling and HLHS who successfully had staged Fontan completion. Because confluence and absence of distortion in the branch pulmonary arteries are essential for successful Fontan completion, PA sling exerts a much greater impact on patients with a single-ventricle physiology than those with a biventricular physiology. There is no question that a symptomatic neonate with PA sling should be repaired at the time of diagnosis even if any kind of cardiac anomalies are associated; however, the fundamental question raised in this patient was whether asymptomatic PA sling should be repaired, and if necessary, the timing of the repair.

PA sling in the present case was untreated for the following reasons. First, this neonate had a small body weight, less than 2.0 kg, which posed technical difficulty as well as risks of late stenosis/occlusion in the implanted left PA. In fact, PA sling repaired in neonatal or early infantile period results in high stenosis or occlusion rate.[3,4] Second, because the main PA was used for aortic arch reconstruction, there was no appropriate site to reimplant the left PA. Finally, concomitant Norwood procedure and PA sling repair might result in prolonged CPB and ischemic time, exposing the patient to risks.

Concerns over untreated PA sling in single-ventricle physiology include turbulence or stenosis/occlusion and subsequent energy loss in Fontan circulation; however, the Fontan circulation has successfully been sustained over 4 years with the evidence of reasonable left PA growth and acceptable pulmonary blood flow distribution. In addition, it is possible that the use of RV-PA shunt as well as the bidirectional BDG tends to provide a symmetric growth pattern of branch pulmonary arteries, and the presence of an additional pulmonary blood flow through the RV-PA shunt after BDG might have, in part, played some roles to facilitate left PA growth in this patient.[5]

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surgery provided tissue stabilization adjacent to the infundibulum, prevented pseudoaneurysm expansion, and minimized risk of rupture. We hypothesize that low right ventricular pressures and containment of the pseudoaneurysm by scar tissue from prior cardiac surgery allowed time for healing and obviated the need for surgery.

## CONCLUSIONS

In selected patients with imaging studies strongly suggestive of traumatic RV pseudoaneurysms and a history of sternotomy, nonoperative management with close monitoring may be considered in the absence of tamponade, increase in pericardial effusion, severe pulmonary hypertension, deterioration in hemodynamic status, or expansion of the suspicious findings on serial imaging studies.

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# Successful Fontan completion in a patient with noncompaction myocardium


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Noncompaction myocardium is a rare cardiomyopathy that represents numerous prominent trabeculations and deep intertrabecular recesses mainly in the left ventricle (LV).[1] The cause of noncompaction myocardium is thought to be abnormal cessation of endomyocardial morphogenesis. Although noncompaction myocardium has been commonly described in association with other structural heart abnormalities,[2] few operative outcomes have been reported. We describe a successful surgical case of tricuspid atresia with noncompaction myocardium.

## CLINICAL SUMMARY

A 22-year-old man was referred to the Okayama University Graduate School of Medicine, Dentistry, and Pharmaceutical Sciences for dyspnea on exertion and progressive cyanosis (oxygen saturation [$S_{PO_2}$] 75% on room air, New York Heart Association II). Tricuspid atresia with normally related great vessels, pulmonary stenosis, and noncompaction myocardium had been diagnosed in the patient at birth. On admission, the patient was active (156 cm, 33 kg) with a normal cardiothoracic ratio and sinus rhythm with incomplete left bundle branch block. Echocardiogram showed spongiform myocardium only in the LV myocardium, large atrial and ventricular communications, and an ejection fraction of 40%. Angiogram demonstrated a single left coronary artery, markedly protuberant trabeculations, and deep intertrabecular recesses in the whole LV (Figure 1, *A, B*), as observed by computed tomography (Figure 1, *C*). Hemodynamic data showed a mean superior vena cava (SVC) and inferior vena cava (IVC) pressure of 8 mm Hg, a mean pulmonary artery (PA) pressure of 11 mm Hg, PA resistance index of 1.7 Wood unit/$m^2$, LV end-diastolic pressure of 7 mm Hg, and a pulmonary to systemic flow ratio of 0.96. In view of the social activity of this young patient and preserved cardiac function, we decided to conduct a staged-Fontan surgery.

The patient was then scheduled to undergo an elective bidirectional Glenn shunt. Anastomosis between the SVC and the right PA was performed without cardiac arrest, using temporary bypass from the SVC through the right atrium, leaving pulmonary antegrade flow. Weaning from circulatory bypass was uneventful, as was the postoperative course, maintaining an $S_{PO_2}$ in the mid 80s until total cavopulmonary connection.

Two years after the bidirectional Glenn shunt, however, this patient had intracerebral hemorrhage caused by arteriovenous malformation and received an emergency coil embolization. He recovered without physical and mental disability. During admission, incidental Holter electrocardiogram


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Disclosures: None.

Received for publication Sept 27, 2008; accepted for publication Feb 24, 2009; available ahead of print June 12, 2009.



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J Thorac Cardiovasc Surg 2010;139:e85-7

0022-5223/$36.00





doi:10.1016/j.jtcvs.2009.02.056

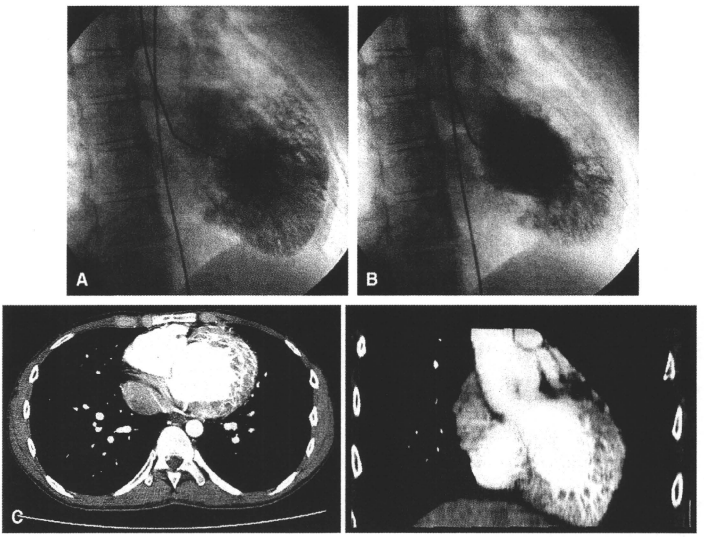


**FIGURE 1.** Prominent thick trabeculations and deep intertrabecular recesses in the whole left ventricular wall are shown by LV angiogram at diastolic (A) and systolic (B) phases (cardiothoracic ratio = 53%). C, Spongiform myocardium depicted by computed tomography.

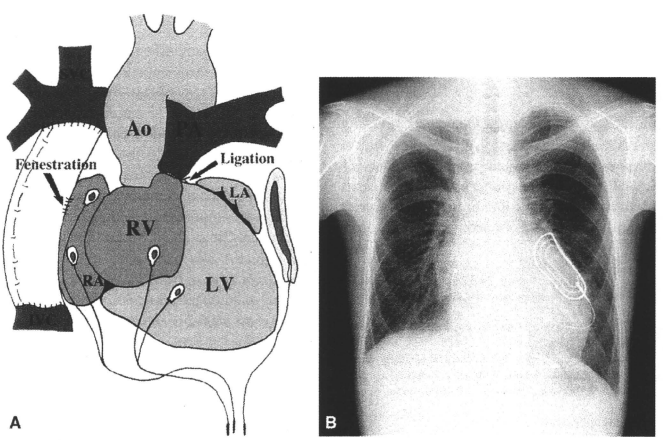


**FIGURE 2.** A, Schema of Fontan operation. B, Postoperative chest x-ray 7 months after Fontan procedure. CTR = 53%. *Ao*, Aorta; *PA*, pulmonary artery; *LV*, left ventricle; *RV*, right ventricle; *LA*, left atrium; *RA*, right atrium; *SVC*, superior vena cava; *IVC*, inferior vena cava.

detected symptomatic nonsustained ventricular tachycardia. Hemodynamic data of the pre-Fontan catheter showed a mean SVC and PA pressure of 9 mm Hg, a mean IVC pressure of 6 mm Hg, a PA resistance index of 1.1 Wood unit/$m^2$, and an LV end-diastolic pressure of 5 mm Hg.

Four years after the bidirectional Glenn shunt and another therapy for atypical mycobacteriosis, total cavopulmonary connection was performed. Cardiopulmonary bypass was established by ascending aorta and bicaval drainage. Extracardiac conduit was interposed between the IVC and the right PA with 4-mm fenestration (Figure 2, *A*). These procedures were also performed without cardiac arrest. Cardiopulmonary bypass was uneventfully weaned with low-dose catecholamine support. Finally, implantable cardioverter defibrillator leads were applied. The patient was extubated in the operating room, and anticoagulant plus antiplatelet therapy was commenced postoperatively. Although fluid leakage from bilateral thoracic cavities continued for 4 weeks, there was no episode of ventricular tachycardia during hospital stay. This patient was discharged on postoperative day 44 with an $S_{PO_2}$ of 94% on room air. Hemodynamic data obtained 1 year after the operation showed a mean SVC, IVC, and PA pressure of 8 mm Hg, a PA resistance index of 1.2 Wood unit/$m^2$, and an LV end-diastolic pressure of 5 mm Hg. Ejection fraction and $S_{PO_2}$ on a patent fenestration flow were 44% and 92%, respectively. During outpatient follow-up, chest radiography showed no signs of heart failure (New York Heart Association I) (Figure 2, *B*).

## DISCUSSION

Noncompaction myocardium is an unclassified cardiomyopathy that is usually associated with other congenital cardiac defects.[1,2] It is well known that the poor clinical prognosis of patients with the isolated form is the result of progressive ventricular dysfunction, a variety of arrhythmia, and systemic thromboembolism.[1,3]

In the present case, our strategy and management to treat this patient were as follows. First, a staged-Fontan surgery achieved ventricular unloading and increased oxygen saturation, avoiding sudden hemodynamic change, for which LV contractility may be conserved. Second, we preserved pulsatile blood flow during cardiopulmonary assist. Although the mechanism of the progressive ventricular dysfunction in noncompaction myocardium has yet to be elucidated, recent reports implicate that coronary malperfusion in the numerous prominent trabeculae is attributable to development of myocardial fibrosis on the basis of pathologic analysis.[4,5] Therefore, avoidance of cardiac arrest may maintain coronary microcirculation. To prevent ventricular tachycardia and systemic thromboembolism, we implanted a cardioverter defibrillator and administered both anticoagulant and antiplatelet agents.

## CONCLUSIONS

Noncompaction myocardium presents a surgical challenge because of its poor clinical prognosis and associated cardiac defects. However, a single ventricle with noncompaction myocardium is not a contraindication for Fontan completion. To our knowledge, this is the first report of a successful Fontan completion for noncompaction myocardium.

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Interactive CardioVascular and Thoracic Surgery

**Anatomical repair of a persistent left superior vena cava into the left atrium**

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*Interact CardioVasc Thorac Surg* 2010;11:199-201; originally published online May 3, 2010;
DOI: 10.1510/icvts.2009.230581

doi:10.1510/icvts.2009.230581

INTERACTIVE CARDIOVASCULAR AND THORACIC SURGERY

Interactive CardioVascular and Thoracic Surgery 11 (2010) 199-201

www.icvts.org

Case report - Congenital

# Anatomical repair of a persistent left superior vena cava into the left atrium

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Received 20 December 2009; received in revised form 5 April 2010; accepted 13 April 2010

**Abstract**

The anatomy of a persistent left superior vena cava (SVC) to the left atrium (LA) without the innominate vein can make it challenging to complete intracardiac repair. We reviewed our five cases of the direct end-to-side anastomosis of SVCs to facilitate anatomical repair of SVC-right atrial connection for biventricular repair. Diagnoses were two partial atrioventricular septal defect with left isomerism, one complete atrioventricular septal defect (CAVSD) with left isomerism, one CAVSD without isomerism and one atrioventricular discordance and double outlet right ventricle with right isomerism. Mean age at the operation was 20±23 months (4–58 months) and body weight was 7.8±3.4 kg (4.8–12.7 kg). After completion of intracardiac repair, the SVC to LA was divided and end-to-side anastomosed to the SVC to the right atrium during cardiopulmonary bypass. No early or late death occurred during follow-up of 14.4±6.9 months (7–23 months). None of the patients developed an obstruction at the anastomosis site of the SVCs. The direct end-to-side anastomosis of SVCs achieved an excellent anatomical SVC–right atrium connection in complex congenital heart diseases.


*Keywords:* Congenital heart disease; Pediatric

## 1. Introduction

Persistent left superior vena cava (SVC) that connects directly with the left atrium (LA) can result in a significant intracardiac shunt. If there is no connecting vein between the two SVCs, surgical repair is generally performed by intracardiac rerouting using a baffle to divert flow from the left SVC to the right atrium [1]. In intra-atrial rerouting, the anatomy between the orifices of systemic veins and pulmonary veins can render the procedure to correct the lesion more complex, thus causing a disturbed venous flow [2–4]. There have been few reports describing extracardiac correction for directly connecting SVCs. We herein review our cases involving direct end-to-side anastomosis of SVCs to repair a SVC–left atrial connection for biventricular repair.

## 2. Case report

From July 2007 to December 2009, we performed the direct end-to-side anastomosis of SVCs in five patients (Table 1). The diagnoses were two partial atrioventricular septal defect with left isomerism, one complete atrioventricular septal defect (CAVSD) with left isomerism, one CAVSD without isomerism and one atrioventricular discordance and double outlet right ventricle with right isomerism. Mean age at the operation was 20±23 months (4–58 months) and body weight was 7.8±3.4 kg (4.8–12.7 kg). Computed tomography revealed that the mean distance between the SVCs at the level of the orifice of the brachiocephalic artery was 32.3±5.5 mm (25.8–41 mm) and the mean size of right SVCs was 6.9±2.3 mm (5.2–10.9 mm) and that of left SVCs was 6.5±2.0 mm (4.9–10.0 mm).

Before cardiopulmonary bypass, the persistent left SVC to the LA was mobilized well and its length was measured to assess the possibility of extracardiac redirection. Cardiopulmonary bypass was established with cannulation to the aortic root, the SVC to the right atrium and the inferior vena cava. After the correction of intracardiac defects, the occlusion of the left SVC showed a significant rise in the venous pressure (>30 mmHg) and we abandoned the option of the simple ligation of the persistent left SVC. The left SVC was divided at the SVC–atrial junction and was end-to-side anastomosed to the right SVC superior to the aortic arch (akin to a new innominate vein) during cardiopulmonary bypass. Postoperative anticoagulant therapy was administered for eight weeks after surgery.

No early and late death occurred during mean follow-up of 14.4±6.9 months (7–23 months). No children experienced disturbed flow or obstruction at anastomosis sites by transthoracic echocardiogram. A catheter examination in patient 1 one year after surgery demonstrated no pressure gradient and a smooth venous flow in the anastomosis site of SVCs (Fig. 1).

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*E-mail address:* uga-strah@hotmail.co.jp (S. Ugaki).

Table 1. Patients' demographics

| No. | Diagnosis | Age (months) | Body weight (kg) | Distance between SVCs (mm) | Right SVC size (mm) | Left SVC size (mm) | Operation |
|---|---|---|---|---|---|---|---|
| 1 | CAVSD(A), PLSVC to LA | 8 | 5.4 | 25.8 | 5.5 | 4.9 | Modified one patch repair, direct anastomosis of SVCs |
| 2 | Dextrocardia, partial AVSD, PRSVC to LA, interrupted IVC and hemiazygos connection, right aortic arch, left isomerism | 24 | 10.2 | 41.0 | 6.8 | 6.0 | Patch closure of ASD, cleft closure of mitral valve, direct anastomosis of SVCs |
| 3 | CAVSD(A), PLSVC to LA, interrupted IVC and azygos connection, left isomerism | 4 | 6.1 | 31.2 | 5.9 | 6.4 | Modified one patch repair, direct anastomosis of SVCs |
| 4 | DORV, PS, PLSVC to LA, right isomerism, s/p Bil.mBTS | 58 | 12.7 | 33.1 | 10.9 | 10.0 | VSD patch closure, atrial septation, RVOTR, direct anastomosis of SVCs |
| 5 | Partial AVSD, PLSV to LA, MR, left isomerism | 6 | 4.8 | 30.5 | 5.2 | 5.4 | Patch closure of ASD, cleft closure of mitral valve, direct anastomosis of SVCs |

SVC, superior vena cava; CAVSD, complete atrioventricular septal defect; PLSVC, persistent left superior vena cava; LA, left atrium; AVSD, atrioventricular septal defect; PRSVC, persistent right superior vena cava; IVC, inferior vena cava; ASD, atrial septal defect; DORV, double outlet right ventricle; PS, pulmonary stenosis; Bil.mBTS, bilateral modified Blalock–Taussig shunt; VSD, ventricular septal defect; RVOTR, right ventricular outflow tract reconstruction; MR, mitral regurgitation.

## 3. Discussion

Persistent left SVC into the pulmonary venous atrium creates a right–left shunt and could cause a brain abscess or cerebral infarction [3]. Several surgical procedures to correct this anomaly have been reported, including ligation of the left SVC, intra-atrial redirection of flow from the left SVC to the right atrium, and reimplantation of the left SVC into the right atrium, pulmonary artery or SVC. Ligation of the vein obliterates the intracardiac shunt, but this procedure is risky unless there are large collateral links in the head that allow unobstructed head and neck venous return into the heart [5]. Reimplantation of the persistent left SVC is preferable, especially when there is a possibility that an intra-atrial baffle may obstruct systemic or pulmonary venous return due to the location of the veins' orifices [2, 3]. In addition, extracardiac procedures can render intracardiac repair easier and shorten cardiac arrest time. The cavopulmonary connection is advantageous in terms of the proximity between the SVC and pulmonary artery [2]; however, high pulmonary artery pressure may well reverse the flow into the left SVC and elevated pulmonary resistance may not permit this connection [3, 4]. The use of a polytetrafluoroethylene graft to connect the SVCs has been successful [3], but this technique is not suitable for small children. Shumacker et al. reported a direct connection into the right atrium [5], but they are commonly located remotely and this anastomosis may have technical difficulties [2]. Reddy et al. also reported a direct reimplantation of the left SVC to the right SVC through a tunnel that had been created between the aortic arch and the pulmonary artery [4]. They thought the persistent left SVC runs through the mediastinum more posterior than the right SVC in most cases and this approach can create a more natural connection. However, creating sufficient space to pass the SVC under the aortic arch may be difficult in some cases due to a dilated aorta or pulmonary artery. Furthermore, this approach may restrict the anastomosis site between the SVCs and cause distortion of the left SVC. Therefore, we chose a route superior to the aortic arch to redirect the flow of the left SVC. We have utilized this procedure since 2007 as the initial choice to repair anomalous venous return to the LA. We have been able to perform this procedure in all cases without technical difficulties. Moreover, all patients experienced good outcomes with no obstruction during follow-up.

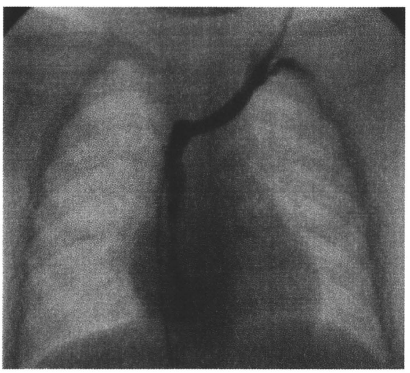

Fig. 1. The catheter study revealed no stenosis at the anastomosis site between the left SVC and the right SVC in the first patient with atrioventricular septal defect. SVC, superior vena cava.

In conclusion, the long-term results of this technique are still unknown, but our findings suggest that the anatomical repair of bilateral SVCs can be an effective treatment strategy for the correction of complex intracardiac anomalies with a persistent left SVC to the LA.

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# Torn Atrial Septum during Transcatheter Closure of Atrial Septal Defect Visualized by Real-Time Three-Dimensional Transesophageal Echocardiography

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Transcatheter closure of atrial septal defects (ASDs) has become an accepted and reliable procedure. Although various complications have been recognized, tear of the atrial septal rim is a rare complication. We report a case of atrial septal rim tear that was diagnosed during the procedure by real-time three-dimensional transesophageal echocardiography (TEE). The device was successfully implanted 3 months after the initial intervention. RT3D TEE is more useful for displaying the entire shape of the defect and its spatial relationship (RT3D) with its neighboring structures compared with conventional two-dimensional echocardiography. By using both two-dimensional and RT3D TEE images, especially in cases with complicated ASD morphology, both the echocardiologist and interventionalist gain valuable information on the morphology of the ASD before and during the procedure. (J Am Soc Echocardiogr 2010;23:1222.e5-1222.e8.)

*Keywords:* Atrial septal defect, Complication, Interventional echocardiography, three-dimensional transesophageal echocardiography

In recent years, transcatheter closure of atrial septal defects (ASDs) has become an accepted and reliable procedure.[1] Tearing of the atrial septum during the procedure is a rare complication, and it is sometimes difficult to make a rapid and accurate diagnosis during the procedure.[2-4]

With the recent rapid developments in technology, real-time three-dimensional (RT3D) transesophageal echocardiography (TEE) with a new fully sampled matrix array transducer has become available.[5] RT3D TEE provides clear and easily understandable en face images. Therefore, this modality has been applied to select patients for transcatheter closure of ASDs and to guide the procedure.[6,7] We report a case of an atrial septum tear during transcatheter closure of an ASD that was visualized by RT3D TEE, allowing us to make an accurate diagnosis during the procedure and to choose an appropriate therapeutic strategy.

## CLINICAL SUMMARY

A 40-year-old woman with exertional dyspnea was referred to the Okayama University Graduate School of Medicine for transcatheter device closure of a secundum ASD. Transthoracic echocardiography showed a left-to-right shunt across the ASD and enlargement in the right atrium and ventricle with no additional associated cardiovascular abnormality.

Transcatheter ASD closure was performed under general anesthesia with the assistance of two-dimensional (2D) TEE, RT3D TEE (Philips IE 33, Philips Medical Systems, Andover, MA), and fluoroscopy. The maximum diameter of the ASD was measured at 18 mm, and an adequate rim around the defect, with the exception of a deficient superoanterior rim, was observed on preprocedural 2D and RT3D TEE. 2D TEE also revealed a thin inferoposterior rim. Pulmonary to systemic flow ratio (Qp/Qs) revealed a significant left-to-right shunt (Qp/Qs ratio 1.93). Balloon sizing with a 34-mm AGA balloon (AGA Medical, Plymouth, MN) resulted in a stretched defect diameter of 20 mm using the stop-flow technique. A 10F AGA sheath was used to deliver the device. A 20-mm Amplatzer septal occluder (ASO) (AGA Medical Corp, Plymouth, MN) was selected on the first attempt. However, the device and delivery sheath easily slipped back into the right atrium, and the device could not be deployed in the proper position. After the first attempt, 2D TEE showed a flailed inferoposterior rim. On the second attempt, a 24-mm ASO was selected, but the device could not be deployed in the proper position. Finally, the maximal longitudinal diameter of the defect was measured at 30 mm. The procedure was terminated, and the ASD could not be closed. Although it was difficult to view the defect in detail using only 2D TEE images, RT3D TEE clearly demonstrated that the borders of the inferior rim were torn and that the defect had increased in size during the procedure (Figure 1). The en face image seen on RT3D TEE made it easier for the operator to evaluate the problem. The inferior rim had been torn when the ASO and delivery sheath slipped back into the atrium, and the defect had become larger during the procedure. The patient was hemodynamically stable during the intervention even after the rim was torn.

After the patient was discharged, no clinical deterioration was observed. The patient was scheduled for surgical closure of the defect,

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0894-7317/$36.00



doi:10.1016/j.echo.2010.04.004

| Abbreviations |
|---|
| **ASD** = Atrial septal defects |
| **ASO** = Amplatzer septal occluder |
| **RT3D** = Real-time three-dimensional |
| **TEE** = Transesophageal echocardiography |
| **2D** = Two-dimensional |

but she refused. Three months after discharge, 2D and RT3D TEE did not reveal flailed surrounding rims, and the torn rim seemed healed, resulting in a deficiency of the superoanterior and inferoposterior rims. The maximal diameter of the defect was 30 mm (Figure 2). Therefore, transcatheter closure of ASD was attempted again.

Right-sided catheterization was performed before the procedure, showing a normal pulmonary artery systolic pressure of 15 mm Hg. The Qp/Qs ratio had increased to 2.88. A 32-mm ASO was selected. The device was successfully implanted, and the ASD was occluded (Figure 3). The patient's exertional dyspnea was improved 1 month after device closure of the ASD. At 1-year follow-up, the patient was still asymptomatic and had no complications (eg, device dislocation).

## DISCUSSION

Tearing of the rim is a rare complication in ASD closure and has been reported in only 3 cases.[2-4] In all these reports, tearing occurred during balloon sizing. In our patient, the inferior rim was very thin, and tearing occurred when the device and delivery sheath slipped back into the right atrium despite gentle manipulation of the catheter. Moreover, the rim was also torn during the second attempt to deploy the device. Therefore, we abandoned the procedure. In previous case reports, surgical intervention was performed in 2 cases with this complication, and 1 was closed with an ASO. Initially, the patient was not scheduled for defect closure with an ASO after the first attempt for fear of device dislocation because of the torn flailing rim. However, the torn rim appeared to be healed 3 months after the first procedure. Therefore, we attempted to close the defect with an ASO, and the procedure was successful. In hindsight, because the thin rim tissue was torn, the final ASO was deployed on the more solid rim tissue, resulting in the stable placement of the ASO. Morphologic variations of ASDs are common, and the surrounding rims include a thin floppy rim,

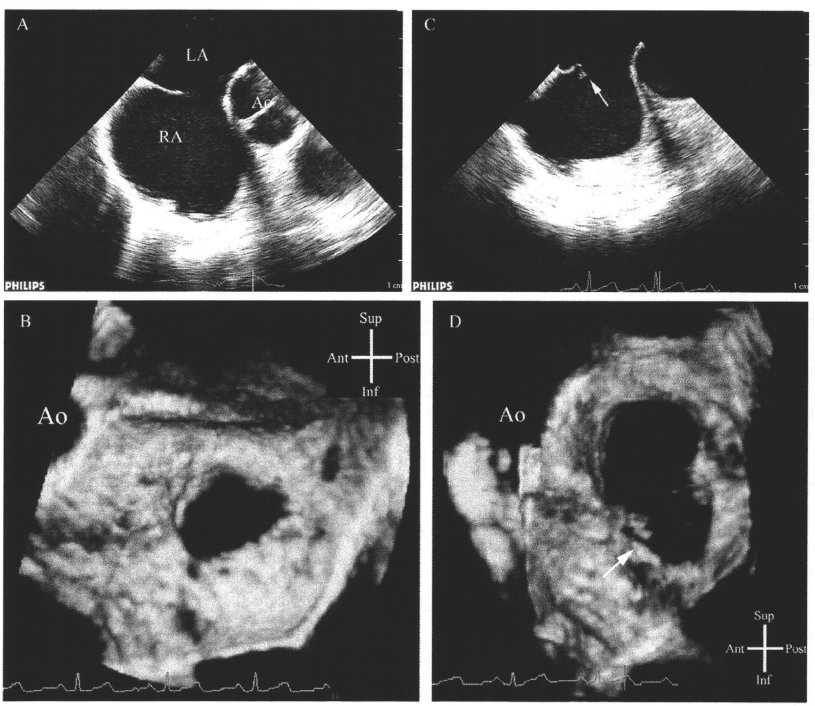


Figure 1 2D TEE **(A, C)** and RT3D TEE **(B, D)** images during transcatheter closure of ASD. **A, B,** The patient initially had a deficient superoanterior rim. **C, D,** After the first attempt, 2D TEE showed a flailed inferoposterior rim (*arrow*). The rim was torn along with the following attempt to deploy the device, and the maximal longitudinal diameter of the defect was measured at 30 mm. *Ao*, Aorta; *LA*, left atrium; *RA*, right atrium; *sup*, superior; *inf*, inferior; *ant*, anterior; *post*, posterior.