Complete Atrioventricular Septal Defect Associated With Tetralogy of Fallot — Surgical Indications and Results —

Yoshihiro Okada, MD; Katsuhiko Tatsuno, PhD; Toshio Kikuchi, MD;
Yukihiro Takahashi, MD; Tomoki Shimokawa, MD

Complete atrioventricular septal defect (AVSD) associated with tetralogy of Fallot is a rare condition that still has problems in the postoperative period. The authors report their surgical experiences over the past 10 years. Nine children underwent total correction. The defect was repaired by the 2-patch technique and the atrioventricular valve was reconstructed by suturing the cleft and annuloplasty. A transannular right ventricular outflow patch was used in 5 patients. All patients had Down syndrome and a free-floating superior bridging leaflet. One patient died from cardiac failure. Although there was no reoperation or death in the late postoperative periods, mild mitral regurgitation occurred in 4 patients and there was moderate or severe pulmonary regurgitation in 2 patients. All survivors currently have no critical symptoms in their daily lives. With the standard of patient selection used, the optimal body weight was around 8 kg and PA index was 200 or more. Right ventriculotomy provided a better view for complete closure of the ventricular septal defect (VSD). In order to avoid re-regurgitation of the atrioventricular valve, the 2-patch technique is the most suitable procedure for total repair. (*Jpn Circ J* 1999; 63: 889–892)

Key Words: Complete atrioventricular septal defect; Surgical management; Tetralogy of Fallot

The combination of complete atrioventricular septal defect (AVSD) and tetralogy of Fallot (TOF) is infrequently observed. Although the mortality of total correction was high in early reports,^{1–4} recent operative outcomes have much improved because of advances in anatomical and clinical recognition of this complex condition.^{5–11} However, postoperative complications such as ventricular septal defect (VSD) recanalization and atrioventricular valvular regurgitation (AVVR) remain high, and some require reoperation. We review our surgical experiences over the past 10 years and the long-term results of this disease, and discuss the relevant surgical management of this lesion.

Methods

Patients and Preoperative Examinations

From 1986 to 1996, 9 children underwent total correction of AVSD associated with TOF. In the same period, we had 2 other cases of this condition who were followed up after palliation. Patient characteristics are presented in Table 1. The study group included 5 boys and 4 girls and their age at the time of operation ranged from 2 to 8 (4.3 ± 2.6) years. Their weight ranged from 8.8 to 21.0 (14.2 ± 5.1) kg. All patients had Down syndrome. Six patients had had one or more previous Blalock-Taussig shunt operations. In all patients, cyanosis had been noted before 1 year of age and the correct diagnosis was established by

echocardiography and angiocardiology.

Electrocardiogram showed sinus rhythm and right ventricular hypertrophy in all patients. First-degree atrioventricular (AV) block was found in 1 patient and incomplete right bundle branch block was observed in 5 patients. The QRS axis showed superior deviation (from -30 to -120 degrees, with a mean of -84.2 degrees) in 7 patients, and the other 2 had right axis deviation ($+120$ degrees). One of the latter 2 patients had no interventricular communication below the inferior bridging leaflet.

In echocardiography, the interventricular communication of the inlet septum was noted to extend primarily to the infundibular portion. All patients had a free-floating superior bridging leaflet of the common AV valve (Rastelli type C). In one case, the grade of aortic overriding was more than 50% (DORV). Although one patient had no AV valve regurgitation, 2 had trivial, 4 had moderate, and 1 had severe regurgitation.

Catheterization data are summarized in Table 2. Systolic pressure in the right ventricle was same as that in the left ventricle. PA indices (PAI)¹² of all patients were over 200 at the time of total repair. There was no case with remarkable left ventricular hypoplasia at the time of total correction.

Operation

Total correction was performed by using cardiopulmonary bypass with moderate hypothermia and cardiac arrest with cold blood crystalloid cardioplegia. The AVSD was repaired by the 2-patch technique. Through a right atriotomy, a comma-shaped patch made of porcine pericardium and reinforced by a Dacron velour patch was sutured on the VSD with interrupted mattress stitches. In 4 cases, the subaortic portion of the defect was closed through a right ventriculotomy. The interatrial communica-

(Received February 19, 1999; revised manuscript received August 13, 1999; accepted August 16, 1999)

Department of Surgery, Sakakibara Heart Institute, Tokyo, Japan
Mailing address: Yoshihiro Okada, MD, 1st Department of Surgery, Chiba University Hospital, 1-8-1 Inohana, Chuoh-ku, Chiba city, Chiba 260-8677, Japan. E-mail: hiro3@x.email.ne.jp

Table 1 Preoperative Patient Characteristics

Case no.	Sex	Age (years)	Other anomalies	Palliation	Rastelli type	AVVR	PAI
1	M	8	PLSVC	Lt. BTA	C	None	297
2	F	2		C	Severe	284	
3	M	2		Bil. BTA	C	Moderate	843
4	M	5	PLSVC		C	Trivial	213
5	M	5		Bil. BTA	C	Moderate	240
6	M	3		Lt. BTA	C	Moderate	394
7	F	5	PLSVC	Bil. BTA	C	Moderate	347
8	F	8		Lt. BTA	C	Mild	360
9	F	1		Lt. single coronary		C	Trivial

All patients have Down syndrome.

PLSVC, persistent left superior vena cava; BTA, Blalock-Taussig anastomosis; PAI, PA index.

Table 2 Preoperative Hemodynamic Data

	Mean	SD	Range
Systolic LVP (mmHg)	93.0	10.0	80–110
Systolic RVP (mmHg)	92.8	10.7	80–110
Systolic PAP (mmHg)	25.8	3.8	24–30
Mean PAP (mmHg)	19.0	3.1	15–22
PA index	377	187	213–843
Sat. O ₂ in aorta (%)	80.1	6.8	67–90
PaO ₂ (mmHg)	47.7	8.6	35–58

LVP, left ventricular pressure; PAP, pulmonary arterial pressure; RVP, right ventricular pressure.

Table 3 Intraoperative Measurements After Repair

Transannular patch	No. patients	Systolic RVP (mmHg)	RVP/SAP
No	4	42.5 (26–56)	0.60 (0.40–0.79)*
Yes	5	48.6 (33–58)	0.64 (0.43–0.94)*
Monocusp patch	3	45.5 (33–58)	0.61 (0.54–0.71)
Nonvalve patch	2	50.7 (44–55)	0.69 (0.43–0.94)
Total	9	45.9 (33–58)	0.62 (0.40–0.94)

*Non-significant. RVP, right ventricular pressure; SAP, systemic arterial pressure.

Table 4 Postoperative Hemodynamic Data

	Mean	SD	Range
Systolic RVP (mmHg)	44.7	9.9	32–60
Systolic PAP (mmHg)	35.6	11.7	20–56
Mean PAP (mmHg)	19.0	3.4	15–25
RVP/SAP	0.46	0.1	0.37–0.61
Cardiac index (L min ⁻¹ BSA ⁻¹)	3.06	0.61	2.3–3.8
PaO ₂ (mmHg)	61.0	8.0	52–73

Postoperative catheterizations were performed from 1 to 3 years after operation. PAP, pulmonary arterial pressure; RVP, right ventricular pressure; SAP, systemic arterial pressure.

tion was closed with a polytetrafluoroethylene patch, which introduced the coronary sinus blood into the right atrial cavity in 7 patients. Repair of the infundibular stenosis was done in all patients. Four patients had a pulmonary annulus of sufficient size, and they had a right ventriculotomy and pulmonary arteriotomy. Repair of the infundibular stenosis was done in all patients. Five patients required a transannular patch, a monocusp patch was used in 3 patients and a non-valve patch was used in 2 patients. Reconstruction of the atrioventricular valve was performed after assessing regurgitation by cold saline injection into the left ventricle. The cleft of the left AV valve was sutured from the base to the tip of the free margin in all patients. Three patients required additional Kay's annuloplasty at the commissural

level. The right AV valvular cleft was sutured in 2 patients. Three patients required reconstruction for peripheral pulmonary arterial stenosis.

Results

Major complications were observed in only 1 of the patients during the perioperative period and that patient died on the 14th postoperative day because of multiple organ failure due to low output syndrome after nonvalved transannular patch repair of the right ventricular outflow tract. As autopsy was not performed, further information could not be obtained.

The other 8 patients each had a fairly good early postop-

erative course. All patients had sinus rhythm. The right ventricular-to-systemic arterial pressure ratio (RVP/SAP), measured immediately after the cardiopulmonary bypass, showed no significant differences between the transannular patch repair group and the nontransannular patch repair group (Table 3). Two patients (8.8 kg and 20.8 kg) were operated on without blood transfusion (reported previously¹³).

Early postoperative echocardiography showed a small leakage at the patch sutured on the VSD in 4 patients. Five patients had mild residual regurgitation of the left atrioventricular valve, and 3 patients had moderate regurgitation.

Postoperative evaluation was performed in 7 patients from 1 to 3 years after operation (Table 4). On echocardiography, 1 patient had no regurgitation of the left atrioventricular valve, 2 patients had trivial regurgitation, and 4 patients had mild regurgitation. Pulmonary valve regurgitation was detected in all 7 patients; it was trivial or mild in 5 patients, and moderate or severe incompetence was detected in 2 patients. Right AV valve regurgitation varied from none to mild. Four patients revealed insignificant residual VSD: in 2 patients the defect closed spontaneously, and in the other 2 a trivial shunt remained. Cardiac output ranged from 2.30 to 3.75 (mean, 3.06 ± 0.61) L min⁻¹ m⁻². Mean PaO₂ at catheterization was 61.0 mmHg.

All patients were followed up, and the periods ranged from 2 to 10 years (mean, 4.2 years). None of the patients had critical symptoms, late death, or reoperation. Four patients were ultimately freed from medication; the other 4 patients were treated with digitalis and diuretics.

Discussion

The association of AVSD and TOF is rare in congenital cardiac malformation. Kirklin et al reported that TOF was found in about 10% of AVSD¹⁴ and AVSD was seen in about 1% of TOF¹⁵. In Sakakibara Heart Institute, TOF appeared in 14.1% of 64 complete AVSDs, and AVSD was found in 4.6% of 196 TOFs. The clinical and anatomical features of our cases did not significantly differ from those in other reports.^{3,4,14,16} All the patients reported here had Down syndrome and a free-floating superior bridging leaflet (Rastelli Type C). On ECG, 8 patients revealed superior axis, and 2 patients showed inferior axis deviation. One of the latter cases did not have the VSD underneath the inferior bridging leaflet, as reported by Vargas et al.¹⁷ All patients in the present series as well as in other reports, showed cyanosis, except for 1 patient who had high pulmonary blood flow and underwent pulmonary arterial banding⁶.

In the early reports the operative mortality of this combined lesion was relatively high, from 29 to 40%, and total correction was thought to be nearly impossible.¹⁻⁴ Over the last several years, the anatomical features of this lesion have been clarified and the surgical technique has been improved. The latest results have attained an acceptable level, in which the operative mortality ranged from 0 to 11%.⁵⁻⁸ However, postoperative problems that need careful management, including reoperation, still exist.

Malm et al have reported 6 cases of reoperation, including 5 cases of residual VSD.⁸ Other authors have indicated that the main causes of reoperation are progressive AVVR,^{5,6} right ventricular outflow tract obstruction, including pulmonary stenosis,^{6,8,17} and residual interventricular shunt.^{8,14,18}

In addition, there are postoperative complications that do

not require reoperation. Alonso et al indicated that slight AVVR and pulmonary valve regurgitation are relatively common postoperative findings.⁵ We also had various dysfunctions after total correction. In the present series, however, none of the patients had a complication serious enough to require reoperation. We believe that careful selection of both patient and operative procedure is essential to eliminate serious postoperative complications.

Our standards of patient selection are similar to those of recent reports.^{5,6,8} The optimal criteria, we believe from our experience, are around 8 kg of body weight and 200 or more of PAI. This condition has potentially triple valvular disease whereas AVSD or TOF alone has either double or single valvular deficiency. It might be expected that this malformation cannot withstand severe hemodynamics, so we apply the criteria more strictly than for AVSD or TOF alone. Karl also emphasizes the importance of palliation.¹⁸ If the weight or PAI fell below these criteria, we would use the systemic-pulmonary shunt operation to grow the pulmonary artery and improve the physical constitution. Performing total correction with these criteria, the mean RVP/SAP immediately after repair was 0.62, and we had no trouble throughout the perioperative period in our series except for 1 patient.

Although Malm et al used an approach beginning from both the right atrium and the pulmonary artery to avoid ventriculotomy, they had a fairly high incidence of residual VSD.⁸ On the other hand, Vouhe et al reported that the combined right-atrial and right-ventricular approach could give satisfactory exposure of the VSD and achieve a good result.¹⁹ We believe that right ventriculotomy provides a better view for complete closure of the VSD, preventing recanalization of the VSD.

In order to avoid recanalization of VSD and recurrence of regurgitation of the atrioventricular valve, the 2-patch technique without incision of the bridging leaflets is thought to be the most suitable procedure for total repair. Trusler et al introduced this technique for AVSD in 1976²⁰ and it includes suture closure of the atrioventricular valvular cleft and annuloplasty suture at the commissures on demand. Backer et al reported good results from this method in 1995 and they noted that this technique resulted in a low incidence of mitral regurgitation.²¹ The 2-patch technique provides a maximum amount of leaflet tissue as well as a wider coaptation zone. Ilbawi et al advocated this technique for cases of complete AVSD with TOF⁶ and we also had good results using it.

References

1. Fisher RD, Bone DK, Rowe RD, Ott VL: Complete atrioventricular canal associated with tetralogy of Fallot: Clinical experience and operative methods. *J Thorac Cardiovasc Surg* 1975; **70**: 265-271
2. Sridaromont S, Feldt RH, Ritter DG, Davis GD, McGoon DC, Edwards JE: Double-outlet right ventricle associated with persistent common atrioventricular canal. *Circulation* 1975; **52**: 933-942
3. Pacifico AD, Kirklin JW, Barger LM Jr: Repair of complete atrioventricular canal associated with tetralogy of Fallot or double-outlet right ventricle: Report of 10 patients. *Ann Thorac Surg* 1980; **29**: 351-356
4. Uretzky G, Puga FJ, Danielson GK, Feldt RH, Julsrud PR, Seward JB, et al: Complete atrioventricular canal associated with tetralogy of Fallot. *J Thorac Cardiovasc Surg* 1984; **87**: 756-766
5. Alonso J, Núñez P, DeLeón JP, Sánchez PA, Villagrà F, Gómez R, et al: Complete atrioventricular canal and tetralogy of Fallot: surgical management. *Eur J Cardiothorac Surg* 1990; **4**: 297-299
6. Ilbawi M, Cua C, DeLeon S, Muster A, Paul M, Cutilella A, et al: Repair of complete atrioventricular septal defect with tetralogy of Fallot. *Ann Thorac Surg* 1990; **50**: 407-412

7. Gatzoulis MA, Shore D, Yacoub M, Shinebourne EA: Complete atrioventricular septal defect with tetralogy of Fallot: diagnosis and management. *Br Heart J* 1994; **71**: 579–583
8. Malm T, Karl TR, Mee RBB: Transatrial-transpulmonary repair of atrioventricular septal defect with right ventricular outflow tract obstruction. *J Card Surg* 1993; **8**: 622–627
9. Bertolini A, Dalmonte P, Bava GL, Calza G, Lerzo F, Zannini L, et al: Surgical management of complete atrioventricular canal associated with tetralogy of Fallot. *Cardiovasc Surg* 1996; **4**: 299–302
10. Delius RE, Kumar RV, Elliott MJ, Stark J, DeLeval MR: Atrioventricular septal defect and tetralogy of Fallot: a 15-year experience. *Eur J Cardiothorac Surg* 1997; **12**: 171–176
11. Nakata S, Imai Y, Takanashi Y, Kurosawa H, Tezuka K, Nakazawa M, et al: A new method for the quantitative standardization of cross-sectional areas of the pulmonary arteries in congenital heart diseases with decreased pulmonary blood flow. *J Thorac Cardiovasc Surg* 1984; **88**: 610–618
12. Takahashi Y, Tatsuno K, Kikuchi T, Okada O, Shimokawa T, Ishitoya H: Open heart surgery without blood transfusion for complete atrioventricular septal defect associated with tetralogy of Fallot. *J Jpn Assoc Thorac Surg* 1997; **45**: 170–173
13. Bharati S, Kirklin JW, McAllister HA Jr, Lev M: The surgical anatomy of common atrioventricular orifice associated with tetralogy of Fallot, double outlet right ventricle and complete regular transposition. *Circulation* 1980; **61**: 1142–1149
14. Kirklin JW, Blackstone EH, Pacifico AD, Brown RN, Barger LM Jr: Routine primary repair versus 2-stage repair of tetralogy of Fallot. *Circulation* 1979; **60**: 373
15. Arciniegas E, Hakimi H, Farooki ZQ, Green EW: Results of total correction of tetralogy of Fallot with complete atrioventricular canal. *J Thorac Cardiovasc Surg* 1981; **81**: 768–773
16. Vargas FJ, Coto EO, Mayer JE Jr, Jonas RA, Castaneda AR: Complete atrioventricular canal and tetralogy of Fallot: Surgical considerations. *Ann Thorac Surg* 1986; **42**: 258–263
17. He G, Mee RBB: Complete atrioventricular canal associated with tetralogy of Fallot or double-outlet right ventricle and right ventricular outflow tract obstruction: a report of successful surgical treatment. *Ann Thorac Surg* 1986; **41**: 612–615
18. Karl TR: Atrioventricular septal defect with tetralogy of Fallot or double-outlet right ventricle: surgical considerations. *Semin Thorac Cardiovasc Surg* 1997; **9**: 26–34
19. Vouhe PR, Neveux JY: Surgical repair of tetralogy of Fallot with complete atrioventricular canal. *Ann Thorac Surg* 1986; **41**: 342–345
20. Trusler GA. Discussion of: Mills NL, Ochsner JL, King TD: Correction of type C complete atrioventricular canal: surgical considerations. *J Thorac Cardiovasc Surg* 1976; **71**: 20–28
21. Backer CL, Mavroudis C, Alboliras ET, Zales VR: Repair of complete atrioventricular canal defects: Results with the 2-patch technique. *Ann Thorac Surg* 1995; **60**: 530–537