Transannular patching is a valid alternative for tetralogy of Fallot and complete atrioventricular septal defect repair

Gianluca Brancaccio, MD, PhD,[a] Guido Michielon, MD,[a] Sergio Filippelli, MD,[a] Gianluigi Perri, MD,[a] Duccio Di Carlo, MD,[a] Fiore S. Iorio, MD,[a] Gianluca Oricchio, MD,[a] Roberta Iacobelli, MD,[b] Antonio Amodeo, MD,[a] and Roberto M. Di Donato, MD[a]

Objective: We report our experience with repair of tetralogy of Fallot associated with complete atrioventricular septal defect, addressing in particular the need for a pulmonary valve in the right ventricular outflow tract.

Methods: Between 1992 and 2006, 33 children with tetralogy of Fallot and complete atrioventricular septal defect were admitted; 26 had Down's syndrome (79%). Thirty-two children had complete repair (18 primary, 14 staged); of the 15 who received initial palliation, 1 died before complete repair. Right ventricular outflow tract obstruction was relieved by transannular patch in 14 cases (42%), infundibular patch with preservation of the pulmonary valve in 7 (21%), and right ventricle–to–pulmonary artery conduit in 11 (33%).

Results: There were no hospital deaths. Actuarial survival was 96% ± 3.9% at 5 years and 85.9 ± 1.1% at 10 years. Multivariate analysis showed that type of relief of right ventricular outflow tract obstruction did not influence survival ($P = .16$), nor did the choice to use a valved conduit ($P = .82$). Primary correction ($P = .05$) and lower weight at repair ($P = .05$) were associated with higher probability of survival. Mean follow-up was 69.3 ± 5.9 months (range 0.2–282 months). There were 2 late deaths. Overall freedom from reoperation was 69% at 5 years and 38% at 10 years. Right ventricular outflow tract reconstruction without use of a valved conduit allowed a significantly higher freedom from reinterventions ($P < .05$).

Conclusions: Tetralogy of Fallot associated with complete atrioventricular septal defect can be corrected at low risk with favorable intermediate survival. Use of right ventricle–to–pulmonary artery conduit can be avoided in two thirds of patients with no impact on survival, possibly improving overall freedom from reintervention.

The association of tetralogy of Fallot (TOF) with complete atrioventricular septal defect (CAVSD) is rare[1,2]; it was detected in 5% of patients having CAVSD repair and in 1.7% of patients having TOF repair.[2-4] This anatomic complex exhibits combined, though distinctive, morphologic features of CAVSD and TOF.[5] Initial experiences with surgical correction of this anomaly were affected by high mortality,[4,6,7] but technical advances have led to better outcomes.[3,8] However, the need for a valved reconstruction of the right ventricular outflow tract (RVOT) still remains undefined. This report shows the dispensability of a pulmonary valve in TOF-CAVSD repair.

MATERIALS AND METHODS

Between 1992 and 2006, 33 consecutive children with CAVSD associated with TOF were referred to Bambino Gesù Pediatric Hospital, Rome, Italy. Excluded from this analysis were children with unbalanced CAVSDs requiring univentricular repair and those with atrioventricular or ventriculoarterial discordance.

After parental consent was obtained, a retrospective review of all medical records was carried out. Follow-up data were obtained from outpatient clinic records or by contact with the referring physicians. Functional status was assessed according to the New York Heart Association classification. The study was approved by the local Ethics Committee.

Surgical Technique

Repair was accomplished by standard cardiopulmonary bypass with bicaval cannulation, mild to moderate hypothermia (28°C–32°C), and intermittent infusion of antegrade blood cardioplegic solution. All preexisting shunts were divided at initiation of cardiopulmonary bypass.

All children had closure of septal defects using the 2-patch technique. The ventricular patch was made of glutaraldehyde-treated autologous or heterologous pericardium and was shaped like a comma. Closure of the anterosuperior extension of the ventricular septal defect was facilitated by exposure through a right infundibular incision. The cleft of the left atrioventricular valve was closed with multiple interrupted monofilament sutures, and competence of the valve was assessed by filling the left ventricle with cold saline solution.

Closure of the ostium primum atrial septal defect followed repair of the left atrioventricular valve and was also achieved by an autologous or heterologous pericardial patch, always leaving the coronary sinus on the right atrial side.

The RVOT was explored from both the right atrium and the infundibular incision. Resection of obstructive right ventricular muscle bundles was performed. The pulmonary valve was sized by Hegar dilators. If the Z-score of the pulmonary valve was ≤–2, the RVOT reconstruction was accomplished by either transannular patch or valved conduit interposition according to the surgeon's preference. More recently, insertion of a polytetrafluoroethylene (PTFE) monocusp valve was combined in cases of transannular patch reconstruction.

Statistical Analysis

Statistical analysis was conducted with the SAS-Statview-1998 statistical software, SAS Institute Inc (Cary, NC). Chi-square analysis was used to compare discrete variables between the groups of children who had staged repair and primary repair of CAVSD-TOF. Continuous variables, expressed

From the Departments of Pediatric Cardiothoracic Surgery[a] and Cardiology,[b] Bambino Gesù Pediatric Hospital, Rome, Italy.

Received for publication Feb 14, 2008; revisions received April 6, 2008; accepted for publication Sept 30, 2008.

Address for reprints: Gianluca Brancaccio, MD, PhD, Dipartimento Medico-Chirurgico di Cardiologia Pediatrica, Ospedale Pediatrico Bambino Gesù, Piazza S. Onofrio, 4, 00165 Roma, Italy (E-mail: gbrancaccio70@hotmail.com).

J Thorac Cardiovasc Surg 2009;137:919-23
0022-5223/$36.00

doi:10.1016/j.jtcvs.2008.09.055

Abbreviations and Acronyms

CAVSD = complete atrioventricular septal defect
PTFE = polytetrafluoroethylene
RVOT = right ventricular outflow tract
TOF = tetralogy of Fallot

as means ± standard deviations, were compared by unpaired *t* testing. One patient (No. 11) died before complete correction and was excluded from the analysis. Categorical analysis was conducted by chi-square and Fisher exact test. Freedom from time-related events was conducted according to actuarial and/or Kaplan-Meier technique; the resulting curves with 95% confidence limits were compared with log-rank testing, and nomograms of the hazard function were obtained. Selected and separate end points were defined as death and reoperation. Early mortality was defined as death within 30 days from surgery or prior to hospital discharge. Variables associated with an increased risk of death and reoperation were assessed by multivariate logistic regression and Cox proportional risk analysis.

RESULTS

Preoperative Data

Among the 33 children admitted to our center, there were 17 boys and 16 girls, with a mean weight of 15.7 ± 14.1 kg. Twenty-six (79%) had Down's syndrome, and 1 had Cantrell syndrome. Cyanosis was present in 15 cases (48%). A Rastelli type A CAVSD was diagnosed in 2 children, and all other 31 patients had the classical type C variant. Two patients had right aortic arch, 6 had persistent left superior vena cava to coronary sinus, and 1 had Wolff-Parkinson-White syndrome.

Surgical Strategy

Primary repair was achieved in 18 patients at a mean age of 32 ± 28.3 months, whereas 15 patients (45%) had initial palliation by systemic to pulmonary shunt (12), right ventricular outflow patch (2), or percutaneous balloon dilatation of the pulmonary valve (1). Mean age at palliation was 26.9 ± 33.5 months (range, 1.4–103.7 months). One patient died suddenly at home after systemic-to-pulmonary shunt, prior to complete repair. The remaining 14 patients had secondary complete repair at a mean of 55.1 ± 58.3 months (range, 1.2–210.7 months) after palliation.

Hospital Outcome

On the whole, 32 children had complete repair at a mean age of 54.9 ± 53.9 months (range, 4.6–224.4 months). There were no early deaths. Table 1 compares the preoperative and operative data between primary and staged repair.

Early reoperation was necessary in 6 patients including mediastinal reexploration for bleeding in 2 patients, diaphragm plication in 1, and implantation of a pacemaker for complete heart block in 3 patients (9%; Nos. 10, 12, and 15; Table 2). Patient No. 28 had urgent reoperation for severe aortic insufficiency on postoperative day 5. The noncoronary cusp of the aortic valve was torn and had successful repair by autologous pericardial patch; the aortic valve function of this patient remains normal 2 years postoperatively.

TABLE 1. Comparison of children having primary and staged repair

Variable	Staged repair	Primary repair	*P*
Patients (n)	15	18	.856
Preoperative			
Age (mo)	81.4 ± 66.1	32 ± 28.3	.655
Weight (kg)	22.2 ± 19.5	11.1 ± 5.5	.055
Sex (M/F)	8/7	9/9	.745
Down's syndrome (n)	15	11	.078
Operative			
ECC (min)	331 ± 109.6	297.5 ± 77.5	.543
XCL (min)	174.8 ± 44.7	180 ± 55.3	.944
Transannular patching (n)	5	9	.083
Valved conduit (n)	5	6	.123
Infundibular patching (n)	4	3	.345
ICU stay (d)	7 ± 6.5	6.1 ± 5.8	.566

ECC, Extracorporeal circulation; *XCL,* aortic crossclamping time; *BT,* Blalock-Taussig; *RVOT,* right ventricular outflow tract; *ICU,* intensive care unit.

Follow-up

Follow-up assessment was available for 31 patients who had complete correction (97%). The mean follow-up was 69.3 ± 5.9 months (range, 0.2–282 months). During this period, there were 2 deaths. Patient No. 4 (Table 2) died 20 days after reoperation for residual ventricular septal defect and pulmonary stenosis with dilatation of RVOT. His postoperative course was complicated by mediastinitis and multiple organ failure. The other patient died at home while awaiting conduit replacement (patient No. 1). Both children had Down's syndrome and both had staged repair of TOF-CAVSD.

Actuarial survival was 96% ± 3.9% at 5 years and 85.9% ± 1.1% at 10 years (Figure 1). Among the 29 survivors, 55% were in class I, 38% in class II, 5% in class III, and 2% in class IV.

Role of RVOT Reconstruction

Right ventricular outflow tract obstruction was relieved by a transannular patch in 14 cases (42%), with a PTFE monocusp in 4 cases, by an infundibular patch with preservation of the pulmonary valve in 7 cases (21%), and a right ventricle–to–pulmonary artery valved conduit interposition was used in 11 cases (33%). The type of relief of RVOT did not influence survival ($P = .16$). Use of a prosthetic conduit ($P = .82$) did not affect surgical outcome (Figure 2).

Staged repair ($P = .05$) and higher weight ($P = .05$) at correction were possibly associated with an increased probability of death (Table 3).

Complications and Reoperations

Eight patients (25%) required late reoperations for conduit replacement (3), closure of residual ventricular septal

TABLE 2. Clinical features and follow-up data on 33 patients with AVSD and TOF

Patient No.	Sex	Weight (kg)*	Genetic syndrome	Palliation	Correction (RVOT)	Follow-up
1	F	14	Down's syndrome	BT shunt	Valved conduit	Death
2	M	11	Down's syndrome	—	Transannular patch	Alive
3	F	9.7	Down's syndrome	—	Valved conduit	Alive
4	M	83	Down's syndrome	BT shunt	Infundibular patch	Death
5	M	4	Down's syndrome	—	Transannular patch	Alive
6	F	20	Down's syndrome	BT shunt	Infundibular patch	Alive
7	F	16	Down's syndrome	—	Transannular patch	Alive
8	F	3.5	—	—	Transannular patch	Alive
9	M	6	Down's syndrome	—	Infundibular patch	Alive
10	M	5.1	Down's syndrome	—	Infundibular patch	Alive
11	F	—	Down's syndrome	BT shunt	—	Death before repair
12	F	30	Down's syndrome	BT shunt	Valved conduit	Alive
13	M	20	Down's syndrome	Perc. Dilat.	Transannular patch	Alive
14	M	20	Down's syndrome	—	Infundibular patch	Alive
15	M	9.1	Down's syndrome	—	Transannular patch	Alive
16	M	24	Down's syndrome	BT shunt	Transannular patch	Alive
17	M	15	Down's syndrome	—	Valved conduit	Alive
18	F	13	Down's syndrome	RVOFP	Valved conduit	Alive
19	M	12	Down's syndrome	BT shunt	Infundibular patch	Alive
20	F	7.4	Cantrell syndrome	—	Valved conduit	Alive
21	M	9.7	Down's syndrome	BT shunt	Infundibular patch	Alive
22	M	11.6	Down's syndrome	BT shunt	Transannular patch	Alive
23	F	10	—	—	Transannular patch	Alive
24	M	19.3	—	—	Transannular patch	Alive
25	M	22	Down's syndrome	BT shunt	Valved conduit	Alive
26	F	28	Down's syndrome	BT shunt	Transannular patch	Alive
27	F	16	—	—	Transannular patch	Alive
28	F	8.5	—	—	Transannular patch	Alive
29	F	8.8	Down's syndrome	BT shunt	Transannular patch	Alive
30	F	12	Down's syndrome	—	Valved conduit	Alive
31	F	20	—	—	Valved conduit	Alive
32	M	15	Down's syndrome	RVOFP	Valved conduit	Alive
33	M	6.6	Down's syndrome	—	Valved conduit	Alive

AVSD, Atrioventricular septal defect; *TOF*, tetralogy of Fallot; *BT*, Blalock-Taussig shunt; *Perc. Dilat.*, perctaneous dilatation; *RVOFP*, right ventricular outflow patch. *Weight at correction.

defect (3), mitral valve replacement (1), and tricuspid valve replacement (1). Overall freedom from reoperation after complete repair was 69% at 5 years and 38% at 10 years. Freedom from reoperations after complete repair was 29% after 6 years in the staged repair group and 50.4% at 10 years in the primary repair group ($P = .11$; Figure 3). Differences between the 2 groups were evident, although not statistically significant, probably due to the small size of the cohort. Freedom from reinterventions at 10 years was 62.2% ± 20% in the no-conduit group compared with 14.0% ± 12.9% in the conduit group ($P = .05$; Figure 4). Presence of trisomy 21 was an inverse risk factor for reintervention according to Cox proportional hazard ($P = .008$; Table 4).

DISCUSSION

Tetralogy of fallot associated with CAVSD is an uncommon finding.[1,2] The current literature reports few studies with small cohorts.[3,5,8-11] To our knowledge, our experience represents one of the largest cohorts with long-term follow-up.

The association of TOF and CAVSD was long considered a surgical challenge.[5,8-11] Mortality rate at correction ranged between 15% and 33%.[10,12,13] Recent reports confirm

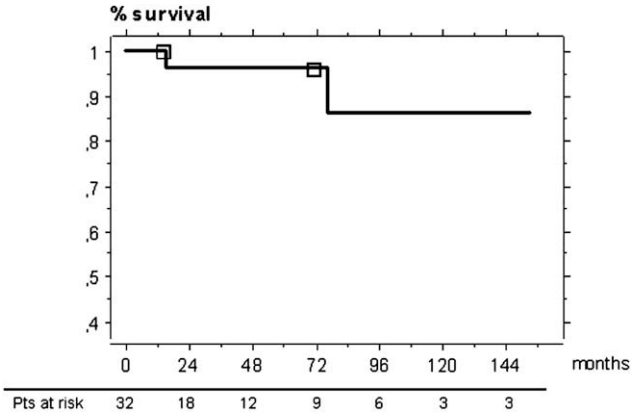

FIGURE 1. Overall actuarial survival after complete repair of tetralogy of Fallot with complete atrioventricular septal defect.

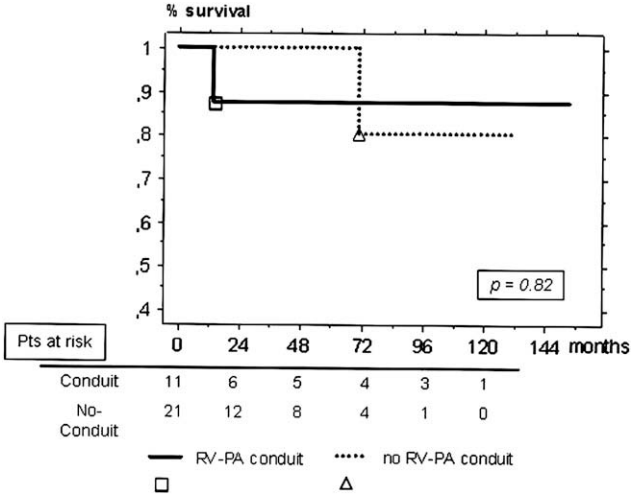

FIGURE 2. Kaplan-Meier survival probability according to type of right ventricular outflow tract repair. *RV-PA*, Right ventricle to pulmonary artery.

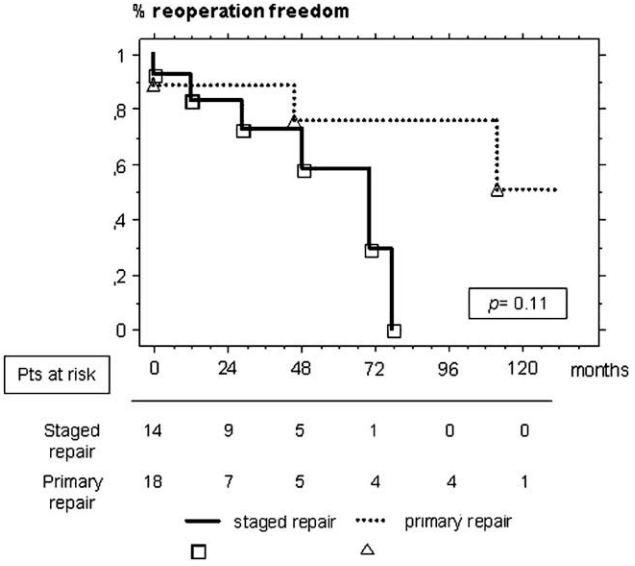

FIGURE 3. Kaplan-Meier freedom from reoperation in primary versus staged repair of tetralogy of Fallot with complete atrioventricular septal defect.

a reduction in mortality and morbidity after surgical correction.[3,5,8] These improvements can be attributed to several factors, including better diagnostic accuracy, perioperative care, and surgical approach.

Our study shows the following:

1. There was no hospital mortality at repair, and the actuarial survival was 86% at 10 years. These findings demonstrate that this anomaly can be corrected with low risk and favorable intermediate survival. Therefore, our data confirm the recent improvements in surgical repair of this complex lesion.[3,5,8,12,13]
2. The type of RVOT obstruction relief did not affect surgical outcome. This finding is in contrast with previous recommendations about the mandatory use of a valved conduit to avoid pulmonary regurgitation particularly in the critical early postoperative period.[10] We suggest that in case of favorable anatomy (normal pulmonary artery size, good right ventricular function), the RVOT obstruction can be effectively relieved by transannular or limited infundibular patching. In our experience, avoidance of right ventricle–to–pulmonary artery conduit was associated with a significantly higher freedom from reintervention. Our current policy includes the construction of a PTFE monocusp valve to reduce early pulmonary regurgitation, yet realizing the short-term efficacy of this approach.[14,15]
3. Primary repair possibly improves long-term survival. This finding is in accordance with the results described by Najm et al,[3] who concluded that primary repair is superior to staged repair. Previous recommendations for

TABLE 3. Forward stepwise Cox proportional hazard model for mortality

Variable	Chi-square	DF	*P*
Female sex	0.186	1	.6660
Age at repair	3.017	1	.0824
Type of repair	0.850	2	.6539
Conduit	0.118	1	.7311
Non-Down's syndrome	0.268	1	.6048
Staged repair	3.571	1	.0588
Weight at repair	3.826	1	.0505
ECC	0.165	1	.6850
XCL	1.287	1	.2566

DF, Degree of freedom; *ECC*, extracorporeal circulation; *XCL*, aortic crossclamping time.

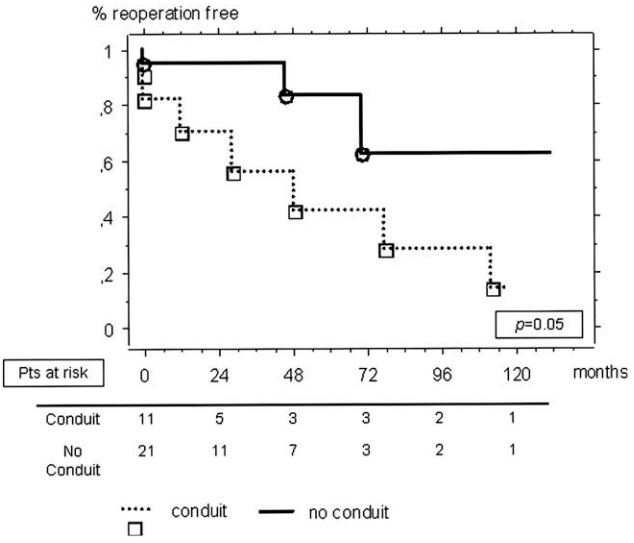

FIGURE 4. Kaplan-Meier freedom from reoperations in conduit and no-conduit group.

TABLE 4. Forward stepwise Cox proportional hazard model for reoperations

Variable	Chi-square	DF	*P*
XCL	0.007	1	.9317
ECC	0.141	1	.7077
Weight at repair	0.243	1	.6220
Primary repair	2.272	1	.1318
Male sex	3.462	1	.0628
Age at repair	0.366	1	.5449
Repair with RV-PA conduit	3.291	1	.0697
Non-Down's syndrome	7.046	1	.0079
Type of repair	3.404	2	.1823

DF, Degree of freedom; *ECC,* extracorporeal circulation; *XCL,* aortic crossclamping time; *RV-PA,* right ventricle–to–pulmonary artery.

age at repair have been in the range of 4 to 6 years[2,4,16]; palliation was endorsed in severely cyanotic infants to carry them through that age target. Our results show that repair at 2 years of age allows excellent outcome. Since 2000, our policy favors primary repair to avoid potential complications (eg, prolonged cyanosis, progressive ventricular hypertrophy, prolonged left ventricular volume overload, interstage mortality, and increasing atrioventricular valve regurgitation).

4. Presence of genetic syndrome was common in our cohort (80%). This finding confirms the recent knowledge about the relationship between trisomy 21 and congenital heart defects.[17,18] However, in contrast to the experience of Karl[8] and Uretzky and colleagues,[4] trisomy 21 was not found to be a risk factor for death. Rather, Down's syndrome appeared a protective factor for freedom from reintervention.

This retrospective study has at least 2 limitations. First, it reports an experience across 15 years, during which important improvements in pre- and postoperative care have been achieved. Second, the small number of patients in some subgroups suggests a careful interpretation of data analysis and requires further confirmation.

CONCLUSION

Teratology of fallot-CAVSD can be corrected at low risk and good intermediate survival. Our results demonstrate that systematic use of right ventricle–to–pulmonary artery conduit can be avoided at least in cases of favorable anatomy. Transannular patching is a valid alternative for RVOT reconstruction and prevents multiple conduit replacement.

References

1. Najm HK, Coles JG, Endo M, Stephens D, Rebeyka IM, Williams WG, et al. Complete atrioventricular septal defects: results of repair, risk factors, and freedom from reoperation. *Circulation*. 1997;96(9 Suppl):II-5.
2. Arciniegas E, Hakimi M, Farooki ZQ, Green EW. Results of total correction of tetralogy of Fallot with complete atrioventricular canal. *J Thorac Cardiovasc Surg*. 1981;81:768-73.
3. Najm HK, Van Arsdell GS, Watzka S, Hornberger L, Coles JG, Williams WG. Primary repair is superior to initial palliation in children with atrioventricular septal defect and tetralogy of Fallot. *J Thorac Cardiovasc Surg*. 1998;116:905-13.
4. Uretzky G, Puga FJ, Danielson GK, Feldt RH, Julsrud PR, Seward JB, et al. Complete atrioventricular canal associated with tetralogy of Fallot. Morphologic and surgical considerations. *J Thorac Cardiovasc Surg*. 1984;87:756-66.
5. Vargas FJ, Coto EO, Mayer JE Jr, Jonas RA, Castaneda AR. Complete atrioventricular canal and tetralogy of Fallot: surgical considerations. *Ann Thorac Surg*. 1986;42:258-63.
6. Bastos P, De LM, Macartney F, Stark J. Correction of type C atrioventricular canal associated with tetralogy of Fallot. *Thorax*. 1978;33:646-8.
7. Zavanella C, Matsuda H, Subramanian S. Successful correction of a complete form of atrioventricular canal associated with tetralogy of Fallot: case report. *J Thorac Cardiovasc Surg*. 1977;74:195-8.
8. Karl TR. Atrioventricular septal defect with tetralogy of Fallot or double-outlet right ventricle: surgical considerations. *Semin Thorac Cardiovasc Surg*. 1997;9:26-34.
9. Vouhe PR, Neveux JY. Surgical repair of tetralogy of Fallot with complete atrioventricular canal. *Ann Thorac Surg*. 1986;41:342-4.
10. Ilbawi M, Cua C, DeLeon S, Muster A, Paul M, Cutilletta A, et al. Repair of complete atrioventricular septal defect with tetralogy of Fallot. *Ann Thorac Surg*. 1990;50:407-12.
11. Alonso J, Nunez P, Perez de LJ, Sanchez PA, Villagra F, Gomez R, et al. Complete atrioventricular canal and tetralogy of Fallot: surgical management. *Eur J Cardiothorac Surg*. 1990;4:297-9.
12. 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.
13. Prifti E, Crucean A, Bonacchi M, Bernabei M, Luisi VS, Murzi B, et al. Total correction of complete atrioventricular septal defect with tetralogy of Fallot. *J Heart Valve Dis*. 2003;12:640-8.
14. Brown JW, Ruzmetov M, Vijay P, Rodefeld MD, Turrentine MW. Right ventricular outflow tract reconstruction with a polytetrafluoroethylene monocusp valve: a twelve-year experience. *J Thorac Cardiovasc Surg*. 2007;133:1336-43.
15. Stewart RD, Backer CL, Young L, Mavroudis C. Tetralogy of Fallot: results of a pulmonary valve-sparing strategy. *Ann Thorac Surg*. 2005;80:1431-8.
16. Pacifico AD, Kirklin JW, Bargeron 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;29:351-6.
17. Michielon G, Marino B, Formigari R, Gargiulo G, Picchio F, Digilio MC, et al. Genetic syndromes and outcome after surgical correction of tetralogy of Fallot. *Ann Thorac Surg*. 2006;81:968-75.
18. Seifert HA, Howard DL, Silber JH, Jobes DR. Female gender increases the risk of death during hospitalization for pediatric cardiac surgery. *J Thorac Cardiovasc Surg*. 2007;133:668-75.