大韓胸部外科學會誌 第21卷 第1號
Vol. 21, No. 1, February, 1988

Tetralogy of Fallot with Subarterial Ventricular Septal Defect

Sam Hyun Kim , M.D.*, Dong Man Seo, M.D.*, Pyo Won Park, M.D.*, Meong Gun Song, M.D.*, Young Kwan Park, M.D.*, and Yung Kyoon Lee, M.D.*

— **Abstract** —

Tetralogy of Fallot with subarterial ventricular septal defect is known to be relatively common in the Far East. Among the 140 consecutive patients who underwent repair for tetralogy, 15 (10.7%) had this variant form of tetralogy.

Though the degree of pulmonic stenosis seems only mild or moderately severe because of the absence of infundibular septum and relatively large size of pulmonary arteries in this type of tetralogy, it has a special surgical implication that a patch widening of the right ventriculotomy is necessary in every case to avoid the postoperative subpulmonic stenosis.

A morphological and surgical features of tetralogy with subarterial VSD are presented.

A variable degree of anterior and leftward deviation of the infundibular septum, with narrowing of the right ventricular outflow tract (RVOT) and a large subaortic ventricular septal defect (VSD), constitute the classic features in the anatomy of tetralogy of Fallot[1,2]. However, in a small number of cases, the infudndibular septum itself may be absent so that the VSD extends to become subarterial. This variant form of a tetralogy with the doubly committed subarterial VSD is reported not uncommon in the Far East[3,11].

Among the 140 consecutive patients who underwent surgical correction of tetralogy from January, 1986 until August, 1987 at Sejong General Hospital, 15 had this atypical form of tetralogy.

This report describes several clinical, morphological and surgical features of tetralogy with doubly committed subarterial VSD.

* Department of Thoracic and Cardiovascular Surgery, Sejong General Hospital, Sejong Heart Institute, Puchon-Shi, Korea

** Presented at the 8th Asian Congress on Thoracic and Cardiac Surgery, Singapore, November, 1987.

Patients

In order to avoid the unnecessary confusion in the description of the location of ventricular septal defect in tetralogy of Fallot, we reviewed the patients with tetralogy since January, 1986, since when we have adopted the Soto's classification[10] for ventricular septal defects.

We noticed four types of ventricular septal defect in tetralogies, perimembranous, infundibular muscular, subarterial and perimembranous-subarterial.

Both the perimembranous and infundibular muscular VSDs are below the deviated infundibular septum and subaortic in their location. In the perimembranous type, the central fibrous body forms a part of the posterior margin of the ventricular septal defect. In the infundibular muscular type, the ventricular septal defect has a completely muscular rim.

In the subarterial type, the infundibular septum is absent so that the cephalad border of the defect is formed by the conjoined aortic and pulmonary valves and the defect therefore is termed as doubly

committed. The inferior margin of the defect is seperated from the tricuspid valve by the well formed muscular tissue, or the subarterial defect may extend to involve the membranous septum and also have the anatomic features of the perimembranous ventricular septal defect. (Fig.1)

The incidence of types of ventricular septal defect is shown in Table 1. 105 cases (75.0%) had perimembranous defect, 20(14.3%) infundibular muscular, 8(5.7%) subarterial and the remaining 7(5.0%) were perimembranous to subarterial in their location.

These can be devided into two groups according to the presence or absence of the infundibular septum. Group A is the classic tetralogy with the ventricular septal defect in subaortic location and group B is the atypical tetralogy with the defect of doubly committed subarterial location. Group A includes the cases with perimembranous defect or infundibular muscular defect. 89.3% of our total cases belonged to this group. The cases with subarterial or perimembranous-subarterial ventricular septal defect are comprised in group B and they were 10.7% of the total number of cases (Fig. 2).

Among the 140 cases who underwent surgical correction, we experienced eight hospital deaths, indicating an overall hospital mortality rate of 5.7%. About one-third of all patients underwent surgery before th age of two.

Though statistically insignificant, those patients in the group B underwent surgical correction at a slightly higher age than those in the group A (Table 2).

The preoperative hemoglobin lebel in the group A were between 9.3 and 27.0 gm% with the mean of 16.5 gm%. In the group B, they were between 11.1 and 19.1 gm% with the mean of 14.7 gm%.

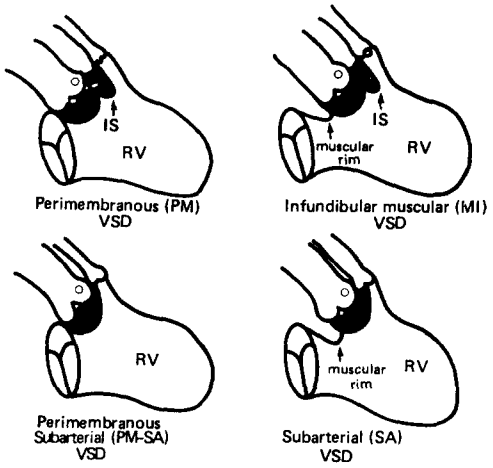

Fig. 1. Schematic Drawings of the four types of VSD in tetralogy

Table 1. Incidence of types of VSD in 140 surgical cases of Tetralogy (January 1986-August, 1987)

Type of VSD	No. ofcase (%)
Perimembranous (PM)	105 (75.0)
Muscular infundibular (MI)	20 (14.3)
Subarterial (SA)	8(5.7)
Perimembranous-Subarterial(PM-SA)	7(5.0)
Total	140 (100.0)

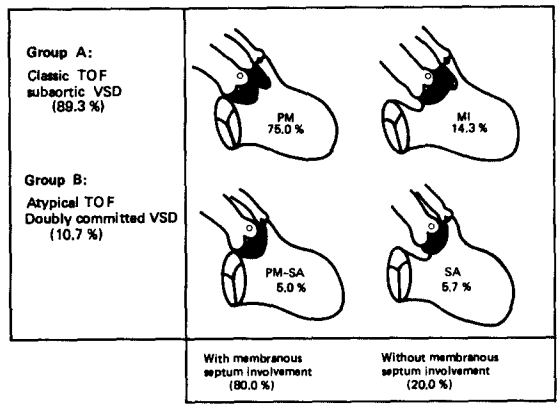

Fig. 2. Location of ventricular septal defects and their relative incidence.

Anatomic considerations

In tetalogy with the subarterial VSD, the infundibular septum was totally absent or had only a hypoplastic fibrous remnant. Therefore the infundibular narrowing was usually mild and mainly formed by the hypertrophied anterior free wall of the right ventricular outflow tract instead of the deviated infundibular septum as seen in the classic tetralogies.

While approximately two-thirds of the group A showed stenosis of the pulmonic valve and required

Table 2. Ages at the time of surgery in two groups and number of hospital death

Age (yr)	Group A		Group B	
	No. of case (%)	No. of HD	No. of case(%)	No. of HD
<2	42 (33.6)	5	2 (13.4)	0
2-10	62 (49.6)	2	7 (46.6)	1
⩾10	21 (16.8)	0	6 (40.0)	0
Total	125 (100.0)	7 (5.6%)	15 (100.0)	1 (6.6%)

HD : hospital death

valvotomy, all cases except one in the group B had stenotic valve. Another significant morphologic difference between the two groups is that of pulmonary arterial lesions requiring surgical intervention. About one-fourth of the group A had pulmonary arterial problems such as segmental branching stenosis or hypoplastic pulmonary arteries, but no one in the group B showed such an unfavorable anatomies. The P value in this relation between the two groups is less than 0.001 (Table 3).

Surgical consideration

In the tetralogies with the doubly committed subarterial VSD, the anatomy of infundibulum and the pulmonary arteries seemed to be favarable for their surgical correction. But as recognized by others previously,[4,5,6] direct closure of the right ventriculotomy would make the apparently good sized infundibulum restrictive.

All cases except one of the tetralogy with the subarterial VSD had the right ventriculotomy widened by a subannular or transannular patch. Transannular patch widening was practised in more than a half of total cases in the group B (Table 4). As all cases in the group B had pulmonary arteries of acceptable size, none need angioplasty of the pulmonary arteries. The one, in whom the right ventriculotomy was closed without widening, died of right heart failure caused by residual subpulmonary stenosis. All survivors are in Functional Class I of the New York Heart Association.

Table 4. Method of RVOT reconstruction

		No. of case (%)
Group A		
	No rt. ventriculotomy	6 (4.8)
	Direct closure	8 (6.4)
	Subannular patch	70 (56.0)
	Transannular patch	41 (32.8)
		125 (100.0)
Groups B		
	Direct closure	1 (6.7)
	Subannular patch	6 (40.0)
	Transannular patch	8 (53.3)
		15 (100.0)

Table 3. Morphology of pumonic flow obstruction

	Stenotic valve	Small annulus	Stenotic PA*
	No. of case (%)	No. of case (%)	No. of case (%)
Group A (n=125)	83 (66.4)	40 (32.0)	32 (25.6)
Group B (n=15)	14 (93.3) p>0.05	6 (40.0) p>0.05	0 (0.0) p<0.001

* Hypoplasia or branch stenosis of pulmonary artery

Table 5. Incidence of postoperative RBBB according to the location of VSD (n=134)

	Type of VSD	No. of cases reviewed	RBBB No. of case (%)
With membranous septum involvement			
	PM	99	74 (74.8)
	PM–SA	7	5 (71.4)
		106	79 (74.5)*
Without membranous septum involvement			
	M1	20	9 (45.0)
	SA	8	2 (25.0)
		28	11 (39.3)*

* Incidence of postoperative RBBB in the cases with or without PM VSD: $p<0.005$

On reviewing the incidence of postoperative right bundle branch block (RBBB), we noticed an interesting finding which may suggest an etiology of postoperative RBBB. Only two (25.0%) of eight tetralogies with the subarterial VSD showed postoperative RBBB, while 74.5% of all tetralogies with VSD of the perimembanous involvement (PM or PM-SA) had right bundle branch block (Table 5). None were complicated with a complete atrioventricular block.

Discussion

Various descriptions has been used for this anatomic variant such as tetralogy of Fallot with absent conus septum[5], tetralogy of Fallot with subpulmonic VSD[5], bulboventricular VSD[15] or subarterial VSD[6,9] This entity is known to be uncommon among the European patients with tetralogy[2,20], but high incidence has been reported from the Middle[4], and the Far East[3,12,16].

The clinical profile in the natural history of these patients is somewhat different from that observed in the classic tetralogy. They are less cyanotic as a group and only a small number of patients need corrective surgery below the age of two years. On preoperative angiography, the degree of pulmonic stenosis seems only mild or moderately severe because the typical subpulmonary bulk usually afforded by the deviated infundibular septum in the classic tetralogy is lacking[9] and the size of the pulmonary arteries are relatively large in almost every case[3,5]. The severity of right ventricular obstruction in these patients depends on the severity of the pulmonic valvular stenosis and on the hypoplasia of the right ventricular outflow tract. All patients except one in our series had valvular stenosis and none had the unfavarable pulmonary arterial problems.

Neirotti and associates[5] and others[4] reported that closure of subarterial VSD in these patients would make an apparently good sized infundibular chamber restrictive and they recommended liberal use of a transannular patch enlargement of the right ventricular outflow tract during the repair. Later, Vargas et al[6] noticed that the transannular patch was frequently uncecessary, and that the patch enlargement confined to the ventriculotomy resulted in the good result. The annulus of the pulmonic valve was nonrestrictive in most of their cases.

In our series, approximately one-half of all tertralogies with the subarterial VSD, which was slightly larger proportion than in those with classic tetralogy, needed the transannular patch widening following the criteria of minimun acceptable pulmonary valve ring diameter propoposed by Pacifico et al[8].

Aortic regurgitation by the prolapse of the right

aortic cusp through the ventricular septal defect were reported[17,18]. Severe dialtation of the aortic root and the turbulent flow up into the dilated aorta may cause the regurgitation in aged patients with tetralogy.

Although the anatomic relationship of the aortic valve to the ventricular septal defect in this from of tetralogy is similar to that in the isolated VSD of the subarterial location, the aortic regurgitation in tetralogy with subarterial VSD has been described to be uncommon[5]. Matsuda[15] et al proposed that blood flow through the VSD in tetralogy is mainly form the right ventricle to the left ventricle, so that the related aortic valve is not likely to be distorted and herniated to result in regurgitation as often seen in the isolated subarterial ventricular septal defect.

REFERENCE

1. Becker AE, Conner M, Anderson RH: *Tetralogy of Fallot. A morphometric and geometric study. Am J Cardiol 35:402, 1975.*
2. Anderson RH, Allowrk SP, Ho SY et al: *Surgical anatomy of tetralogy of Follot. J Thorac Cardiovasc Surg 81:887, 1981.*
3. Ando M : *Subpulmonary ventricular septal defect with pulmonary stenosis (letter). Circulation 50:412, 1974.*
4. Capelli H, Somerville J: *Atypical Fallot's tetralogy with doubly committed subarterial ventricular septal defect. Diagnotic valve of 2-dimensional echocardiography. Am J Cardiol 51:282, 1983.*
5. Neirotti R, Galindez E, Kreutzer G et al: *Tetralogy of Follot with subpulmonary ventricular septal defect. Ann Thorac Surg 25:51, 1978*
6. Vargas FJ, Kreutzer GO, Pedrini M et al: *Tetralogy of Fallot with subarterial ventricular septal defect. Diagnostic and surgical consideration. J Thorac Cardiovase Surg 92:908, 1986.*
7. Alfieri O, Blackstone EH, Kirklin JW et al: *Surgical treatment of tetralogy of Fallot with pulmonary atresia. J Thorac Cardiovasc Surg 76:321, 1978*
8. Pacifico AD, Kiklin JW, Blackstone EH: *Surgical management of pulmonary stenosis in tetralogy of Fallot. J Thorac Cardiovasc Surg 74:382, 1977*
9. Soto B, Pacifico AD, Ceballos R et al: *Tetralogy of Fallot: An angiographic-pathologic correlative study. Circulation 64:558, 1981.*
10. Soto B, Becker AE, Moulaert AJ et al: *Classification of isolated ventricle septal defects. Br Heart J 43:332, 1980.*
11. Lee HJ, Yeum MK, Ko JK et al: *Clinical profile and diagnostic outcome of TOF in Korea. J Sejong Gen Hosp 4:3, 1987*
12. Yeun KM: *Kim JC, Han MC: Cineangiographic analysis of TOF. J Korean Radio Soci 21: 454, 1985.*
13. Hazan E, Bical O, Bex JP et al: *Is right bundle branch block avoidable in surgical correction of tetralogy of Fallot? Circulation 62:852, 1980.*
14. Gelband H, Waldo AL, Kaiser GA et al: *Etiology of right bundle-branch block in patients undergoing total correction of tetralogy of Fallot. Circulation 44:1022, 1971.*
15. Matsuda H, Ihara K, Mori T et al: *Tetralogy of Fallot associated with aortic insufficiency: Ann Thorac Surg 29:529, 1980.*
16. Kozuka T, Nosaki T, Sato K: *Ventricular septal defect in tetralogy of Fallot. Am J Roentgenol 110:497, 1970.*
17. Bahnso HT, Spencer FC, Landtman B et al: *Surgical treatment and fellow-up of 147 cases of tetralogy of Fallot treated by correction. J Thorac Cardiovasc Surg 44:419, 1962*
18. Lev M, Eckner FAO: *The pathologic anatomy of tetralogy of Fallot and its variations. Dis Chest 45:251, 1964.*
19. Kirklin JW, Karp RB: *The tetralogy of Fallot From a surgical view point. Philadelphia, Saunders, 1970, p34*
20. Rosenquist GC, Sweeney L, Stemple D et al: *Ventricular septal defect in tetraogy of Fallot. Am J Cardiology 31:749, 1973*