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Tetralogy of Fallot With Obstruction of the Ventricular Septal Defect: Spectrum of Echocardiographic Findings

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Tetralogy of Fallot with a restrictive ventricular septal defect and suprasystemic right ventricular pressure is an uncommon anomaly with a high mortality rate. In previous studies, the identity of the tissue obstructing the ventricular septal defect has usually not been determined preoperatively. This report describes the echocardiographic and anatomic features in 4 patients with a restrictive ventricular septal defect among 269 patients with tetralogy of Fallot undergoing surgical repair. Echocardiography determined the presence and identity of the obstructing tissue in all four patients. In one patient, the defect was small in association with marked septal hypertrophy. In three patients, accessory or excessive tricuspid valve tissue obstructed the defect in a manner similar to spontaneous closure of isolated membranous ventricular septal defects.

Autopsy and catheterization findings are also presented of an additional case with a unique mechanism of obstruction by a tricuspid valve with Ebstein's anomaly. In patients with tetralogy of Fallot, recognition of an obstructed ventricular septal defect is important because it appears to have a poor prognosis.

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Tetralogy of Fallot is generally characterized by an unrestrictive malalignment-type ventricular septal defect (1). Tetralogy of Fallot with a restrictive ventricular septal defect and suprasystemic right ventricular pressure is an uncommon entity (2–14) with a relatively high operative mortality rate (6–9,12). Postmortem studies (2,4,6,7,9–13) have demonstrated two primary mechanisms for restriction of the size of the ventricular septal defect; the most frequent is obstruction of the usual defect by abnormal tricuspid valve tissue. Less frequently, the defect may be unusually small or appear to be so as a result of obstruction by a hypertrophied septal band (3–6,8,12). Delineation of the pathologic anatomy may be of prognostic importance and may reduce surgical risk (14). The identity of the obstructing tissue has not frequently been determined by angiography before surgery or necropsy (9,11,12). Echocardiographic findings have been reported (14) previously in only one case of tetralogy of Fallot with restrictive ventricular septal defect. This report presents the echocardiographic and anatomic features of four cases of tetralogy of Fallot with restrictive ventricular septal defect and suprasystemic right ventricular pressure.

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Methods

Patient selection. The cardiac catheterization records were reviewed of all patients with unrepaired tetralogy of Fallot undergoing cardiac catheterization at our institution from January 1982 to February 1986. Patients were selected if catheterization and angiography revealed tetralogy of Fallot or tetralogy of Fallot with pulmonary atresia and a right ventricular peak systolic pressure that exceeded the left ventricular or aortic peak systolic pressure by at least 10 mm Hg. One patient (Case 4) was included on the basis of catheterization data from another institution that met the selection criteria and echocardiographic and surgical observations from our hospital. Doppler echocardiographic examination of this patient demonstrated findings consistent with a right ventricular systolic pressure that exceeded left ventricular systolic pressure by approximately 30 mm Hg. Biplane cineangiography of the left ventricle in the long-axis oblique and right anterior oblique projections and of the right ventricle in frontal and lateral projections was performed routinely in all patients with tetralogy of Fallot, including the four reported here.

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Table 1. Summary of Four Cases of Tetralogy of Fallot With Obstructed Ventricular Septal Defect

Case No.	Echocardiography	Cardiac Catheterization	Outcome	Autopsy
1	TF, PA, VAS, LVH, marked small VSD	TF, PA, RV 180/11 mm Hg, LV 165/18 mm Hg, Ao 75/45 mm Hg, small VSD identified	Died 1 day after aortic valvotomy and RVOT patch	TF, PA, VAS, LVH, VSD small in part because of large right posterior division of septal band, TV not obstructive, large mucous plug at carina
2	TF, PA, small RV, redundant SLTV obstructing VSD	TF, PA, RV 105/8 mm Hg, Ao 66/36 mm Hg, no LVOT obstruction, obstructing SLTV identified, absent RCA ostium, RV sinusoidal connections to abnormal PDCA and RCA	Died unexpectedly at 4 days of age from ventricular arrhythmia 1 h after catheterization	TF, PA, small RV, TV appears immature, redundant supermedial infundibular part of SLTV protrudes into VSD, abnormal RCA and PDCA
3	TF, VSD obstructed by Acc TV tissue, marked RVH	TF, RV 105/8 mm Hg, Ao 80/40 mm Hg, no LVOT obstruction, Acc TV tissue not identified	Died intraoperatively at 9 weeks of age	TF, VSD obstructed by Acc TV tissue continuous with and superior to ALTV
4	TF, PA, malalignment VSD obstructed by Acc TV tissue, small muscular VSD with RV to LV shunt at 2.7 m/s by Doppler study consistent with RV systolic pressure greater than LV by 30 mm Hg	TF, PA, RV 76/4 mm Hg, Ao 65/37 mm Hg, no LVOT obstruction, Acc TV tissue not identified	Underwent successful surgical repair at 19 months of age; VSD obstructed by Acc TV tissue noted intraoperatively	

Acc = accessory; ALTV = anterior leaflet of tricuspid valve; LV = left ventricle; LVH = left ventricular hypertrophy; LVOT = left ventricular outflow tract; m/s = meters per second; PA = pulmonary atresia; PDCA = posterior descending coronary artery; RCA = right coronary artery; RV = right ventricle; RVH = right ventricular hypertrophy; RVOT = right ventricular outflow tract; SLTV = septal leaflet of tricuspid valve; TF = tetralogy of Fallot; TV = tricuspid valve; VAS = valvular aortic steno VSD = ventricular septal defect.

Echocardiographic studies. It is routine at our institution for all patients with tetralogy of Fallot to be evaluated preoperatively by echocardiography. All four study patients were examined by two-dimensional echocardiography using either a Diasonics Cardiovue 100 or Hewlett Packard ultrasound imaging system (model 77020 A) before cardiac catheterization. In all patients, multiple imaging views were obtained, including short- and long-axis views from the subxiphoid, parasternal, apical and suprasternal approach. The subxiphoid and parasternal long- and short-axis views were the best views for imaging the ventricular septal defect and tricuspid valve apparatus.

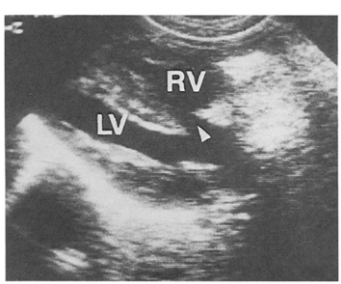

Figure 1. Case 1. Parasternal long-axis echocardiographic view illustrating a small ventricular septal defect (**triangle**) and marked septal hypertrophy. LV = left ventricle; RV = right ventricle.

Direct observations of anatomy. The pathologic anatomy was confirmed by direct visualization in all four patients at surgery or autopsy, or both. Review of the records of the Cardiac Pathology Registry at our institution revealed one additional patient with the diagnosis of tetralogy of Fallot with a restrictive ventricular septal defect who underwent cardiac catheterization in 1961 and died after surgery later the same year.

Results

Incidence and clinical course (Table 1). Between January 1982 and February 1986, 269 patients with unrepaired tetral-

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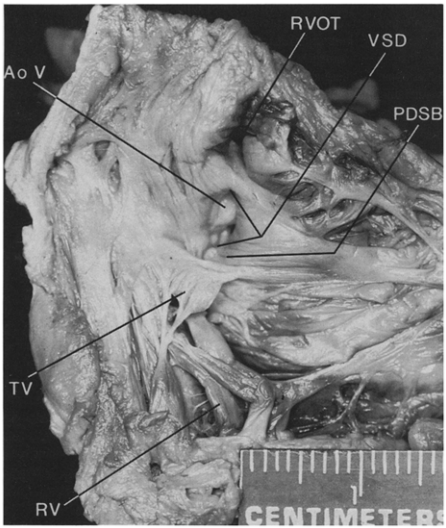


Figure 2. Case 1. View of the opened right ventricle (RV) showing the small ventricular septal defect (VSD). The tricuspid valve (TV) attaches to the right ventricular septal surface normally, near the hypertrophied posterior division of septal band (PDSB). The tiny right ventricular outflow tract (RVOT) becomes atretic distally. The aortic valve (AoV) can just be seen through the ventricular septal defect.

ogy of Fallot (with or without pulmonary atresia) underwent intracardiac surgical repair at our institution. During this time, three patients (Cases 1 to 3) with tetralogy of Fallot undergoing cardiac catheterization at our institution and an additional patient (Case 4) referred for surgery (with catheterization performed elsewhere) were demonstrated to have a restrictive ventricular septal defect and suprasystemic right ventricular pressure. All four patients were studied at our institution by two-dimensional echocardiography and had findings typical for tetralogy of Fallot. Additional defects in these four patients included pulmonary atresia (Cases 1, 2 and 4), multiple ventricular septal defects (Case 4) and valvular aortic stenosis (Case 1).

The perioperative mortality rate for intracardiac repair was considerably higher in the four patients with tetralogy of Fallot with an obstructed ventricular septal defect (3 patients, 75%) than in the 265 patients (16 patients, 6%) with an unobstructed ventricular septal defect (including those with pulmonary atresia, absent pulmonary valve, multiple ventricular septal defects and other associated anomalies). The presence of additional anomalies, particularly pulmonary atresia, may have contributed to the higher mortality rate in patients with an obstructed ventricular septal defect, but did not appear to be wholely responsible. The 30 day surgical mortality rate for intracardiac repair of tetralogy of Fallot in patients with pulmonary atresia and an unrestrictive ventricular septal defect was 15% (7 of 47) in comparison with 67% (2 of 3) in patients with an obstructed ventricular septal defect. The small number of patients with obstructed ventricular septal defect precludes meaningful statistical analysis.

Mechanisms of obstruction of the ventricular septal defect. One patient (Case 1) had tetralogy of Fallot, pulmonary atresia and a severely stenotic bicommissural aortic valve with marked hypertrophy of the left ventricle. Two-dimensional echocardiography showed that the ventricular septal defect was abnormally small, especially during systole (4 mm diameter), as a result of hypertrophy of the ventricular septum along the inferior rim of the defect, particularly the septal band (Fig. 1). The patient died after aortic valvotomy and repair of the right ventricular outflow tract at 1 month of age. At autopsy, the ventricular septal defect was small, in part because of impingement along the inferior border by a prominent right posterior division of the septal

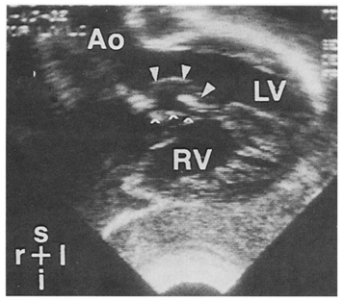


Figure 3. Case 4. Subxiphoid long-axis echocardiographic view of left ventricle (LV) demonstrating accessory tricuspid valve tissue **(solid triangles)** protruding through the ventricular septal defect into the left ventricular outflow tract. The septal leaflet of the tricuspid valve **(open arrows)** can be seen separately in the right ventricle (RV). Ao = aorta; i = inferior; l = left; r = right; s = superior.

band (Fig. 2). Clinical and autopsy findings suggested that a tracheal mucous plug was a significant contributing cause of death.

In three of the four patients, echocardiography demonstrated obstruction of the ventricular septal defect by tricuspid valve tissue, billowing across the ventricular septal defect into the left ventricular outflow tract during systole (similar to a spinnaker sail). This was best visualized in the subxiphoid and parasternal long-axis views. In two of these patients (Cases 3 and 4), the obstruction was caused by a pouch of accessory tricuspid valve tissue, which was superior to and distinct from the three leaflets of the tricuspid valve, but with attachments to the crest of the ventricular septum and the parietal band (Fig. 3). In one patient (Case 4) with serial echocardiographic examinations, the echocardiographic appearance of the accessory tricuspid tissue did not change from 2 days to 19 months of age. Direct observation at surgery (Cases 3 and 4) and autopsy (Case 3) confirmed partial obstruction of the ventricular septal defect by accessory tricuspid valve tissue that was separate from, but widely continuous with, the anterior leaflet of the tricuspid valve and that had chordal attachments to the infundibular septum (Fig. 4).

The ventricular septal defect was obstructed in one patient (Case 2) by an abnormally redundant outpouching of valvelike tissue continuous with the septal leaflet of the tricuspid valve (Fig. 5). At autopsy the tricuspid valve appeared myxomatous and immature. An unusually prominent spinnaker-shaped infundibular leaflet of the tricuspid valve was adjacent to the anterior border of the septal leaflet and protruded into and partially obstructed the ventricular septal defect (Fig. 6). Although the tricuspid valve tissue projected into the left ventricular outflow tract during systole in all three of these patients, in none did this tissue produce any physiologically significant stenosis of the left ventricular outflow tract, as determined by Doppler echocardiography and catheterization.

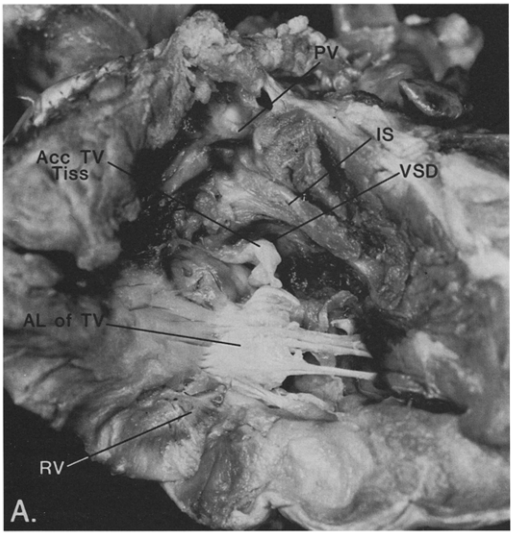
A fifth case (Case 5) is presented because of the unique mechanism responsible for obstruction of the ventricular septal defect. Cardiac catheterization of this male infant in 1961 showed an atrial and a ventricular septal defect with right to left blood flow, right ventricular infundibular stenosis and suprasystemic pressure (peak systolic pressure 95 mm Hg in the right ventricle, 70 mm Hg in the left ventricle). The infant died at 4 months of age after a left Blalock-Taussig shunt procedure. Examination of the heart specimen demonstrated tetralogy of Fallot, Ebstein's anomaly of the tricuspid valve, secundum atrial septal defect and a persistent left superior vena cava connected to the coronary sinus. An unusual outpouching of the malformed septal leaflet of the tricuspid valve protruded into and covered the ventricular septal defect (Fig. 7).

Angiographic findings. Angiography demonstrated the small size of the ventricular septal defect in Case 1 and obstruction of the defect by tricuspid valve tissue in Case 2. In the two patients with accessory tricuspid tissue, either no obstructing tissue could be seen or the nature of the tissue could not be identified by angiography.

Discussion

Incidence. Tetralogy of Fallot with restrictive ventricular septal defect and suprasystemic right ventricular pressure is an uncommon anomaly with a relatively high mortality rate (2–14). Our experience has been similar to that previously reported, with an occurrence of restrictive ventricular septal defect in 4 (1.5%) of 269 patients undergoing intracardiac repair of tetralogy of Fallot (with or without pulmonary atresia) at our institution over a 4 year period. The echocardiographic features were similar to the two basic anatomic patterns seen in the postmortem studies (2–14) of tetralogy of Fallot with obstruction of the ventricular septal defect.

Anatomy. The most common anatomic finding in this series and others (2,4,6,7,9–13) is obstruction of the ventricular septal defect by abnormal or accessory tricuspid valve tissue. The obstructing tissue varied, being superior and separate from the tricuspid valve leaflets in Cases 3 and 4, and adjacent to the anterior portion of the septal leaflet in Case 2. However, all three cases were similar in that the obstructing tissue was located between the septal and anterior leaflets of the tricuspid valve (that is, involving the "infundibular leaflet," also known as the medial portion of



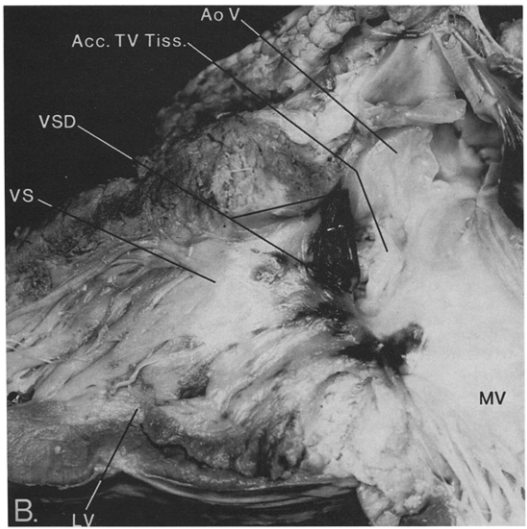


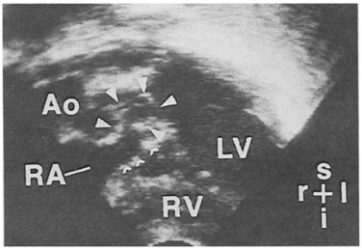
Figure 4. Case 3. **A**, View of the opened right ventricle (RV). The anterior leaflet of the tricuspid valve (AL of TV) can be seen in relation to the accessory tricuspid valve tissue (Acc TV Tiss), which largely obscures the ventricular septal defect (VSD). The narrow right ventricular outflow tract leads to a stenotic pulmonary valve (PV) above the conal or infundibular septum (IS), which also provides one of the attachments of the abnormal tricuspid valve tissue. **B**, View of the opened left ventricle (LV) showing the left ventricular septal surface (VS) with a subaortic malalignment-type ventricular septal defect (VSD), which is partly filled by accessory tricuspid valve tissue (Acc TV Tiss). The aortic valve (AoV) and anterior leaflet of the mitral valve (MV) can be seen in direct fibrous continuity with each other.

the anterior leaflet of the tricuspid valve). Similar pouches or flaps of valve tissue may be seen at the anulus of the tricuspid valve in the anteroseptal commissure of normal hearts (15,16). The echocardiographic findings are very similar to those of a case described by LaCorte et al. (14). Postmortem findings in our Case 5 with tetralogy of Fallot and Ebstein's anomaly demonstrate obstruction of the ventricular septal defect by an unusual myxomatous outpouching of the malformed and displaced septal leaflet (17). Obstruction of an isolated muscular ventricular septal defect by a similar outpouching of the tricuspid valve with Ebstein's anomaly was described by Chesler et al. (18).

The shape, position and attachments of the tricuspid tissue that spontaneously obstructs isolated membranous ventricular septal defects (18–23) are very similar to the tricuspid tissue responsible for obstruction of the malalignment-type ventricular septal defect in three of our patients with tetralogy of Fallot. Similar accessory tricuspid tissue and abnormal valve attachments also occur commonly with malalignment-type ventricular septal defect in association with transposition of the great arteries (24,25). Although obstruction of the left ventricular outflow tract by tricuspid tissue occurs with transposition (25) and other lesions (26), we and others (2–14) have not seen this with tetralogy of Fallot. Rarely, in patients with tetralogy of Fallot, the ventricular septal defect may be obstructed because of anomalous attachments of the mitral valve to the ventricular septum, and in this situation, there is severe left ventricular outflow tract obstruction (27).

The anatomic findings in Case 1 are similar to the second type of obstructed ventricular septal defect described by Fisher et al. (12). The defect was small, in part because of impingement by the right posterior division of the septal band and the very hypertrophied interventricular septum. This represents extreme prominence of the right posterior division of the septal band, which forms the posterior margin of the ventricular septal defect in tetralogy of Fallot (28–30). Previous reports of primary restriction of the size of the ventricular septal defect (3–6,8) and obstruction of the defect by muscle bundles (6) may be the same. The generalized hypertrophy of the interventricular septum in our Case 1 may have been part of the left ventricular hypertrophy associated with valvular aortic stenosis.

Figure 5. Case 2. Subxiphoid long-axis echocardiographic view of the left ventricle (LV) showing a redundant septal leaflet of the tricuspid valve (**solid triangles**) billowing through the ventricular septal defect and into the left ventricular outflow tract. The anterior tricuspid leaflet (**open arrows**) can be seen in the right ventricle (RV). RA = right atrium; other abbreviations as in Figure 3.



Diagnostic studies. Angiography may not distinguish the anatomic basis for aneurysmal obstruction of ventricular septal defect (31–33). Angiography identified the obstructing tissue in only two of our four cases. Echocardiography, however, detected the presence and anatomic mechanism of obstruction of the ventricular septal defect in all four of the four patients with tetralogy of Fallot found at cardiac catheterization to have restrictive ventricular septal defect.

Factors that may influence mortality. Patients with tetralogy of Fallot with an obstructed ventricular septal defect have been recognized previously (6–9,12,14) to have a high mortality rate similar to that seen in our series. Inaccurate preoperative anatomic diagnosis has been proposed as a contributing factor (14). Nevertheless, there appeared to be an elevated mortality rate in our patients (75%) in comparison with our overall experience (of only 6%) with tetralogy of Fallot despite accurate preoperative diagnosis.

Suprasystemic right ventricular pressure in these patients may lead to a greater degree of ventricular hypertrophy, which may have an adverse effect on ventricular compliance (6,34). A severely hypertrophied noncompliant right ventricle may not tolerate the acute diastolic volume load of surgically induced pulmonary regurgitation, thereby adversely affecting surgical outcome (35,36). Consideration should be given to minimizing the width of the transannular patch, if such patching is judged necessary, to decrease pulmonary regurgitation. If a conduit must be placed, it should probably contain a valve.

One patient (Case 2) who died unexpectedly of ventricular tachycardia and fibrillation had absence of the right coronary ostium and abnormal right and posterior descending coronary arteries with sinusoidal connections to the right ventricle. The constellation of a small hypertensive right ventricle, coronary angiopathy and documented ventricular arrythmia is reminiscent of pulmonary atresia with intact ventricular septum (37) and suggests a similar pathophysiologic process involving myocardial ischemia. A recent study (38) demonstrated histologic evidence of myocardial ischemia in hearts with pulmonary atresia and an intact ventricular septum in the absence of coronary artery fistula or dysplasia. Although macroscopic coronary anomalies or myocardial infarcts were not evident in the three other

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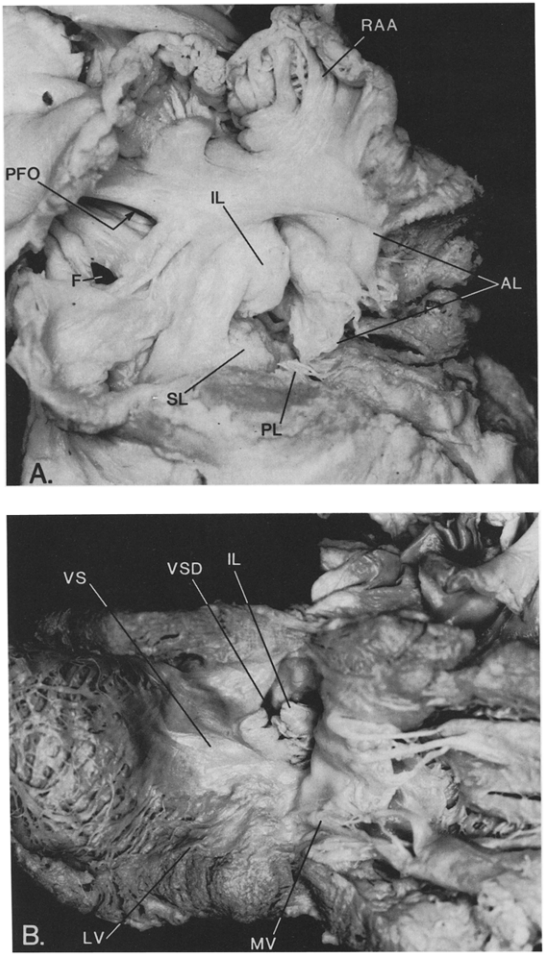

Figure 6. Case 2. A, View of the opened right atrium and tricuspid valve showing a patent foramen ovale (PFO) and a tiny foramen (F) in the substance of the septum primum, the right atrial appendage (RAA), the infundibular leaflet (IL) of the tricuspid valve adjacent to the septal leaflet (SL) of the tricuspid valve, the anterior leaflet (AL) of the tricuspid valve and the small myxomatous posterior leaflet (PL) of the tricuspid valve. B, This view of the opened left ventricle (LV) shows the relation of the infundibular leaflet (IL) of the tricuspid valve to the ventricular septal defect (VSD) and its attachments to the crest of the interventricular septum (VS). The mitral valve (MV) is seen attaching normally to the left ventricular free wall.

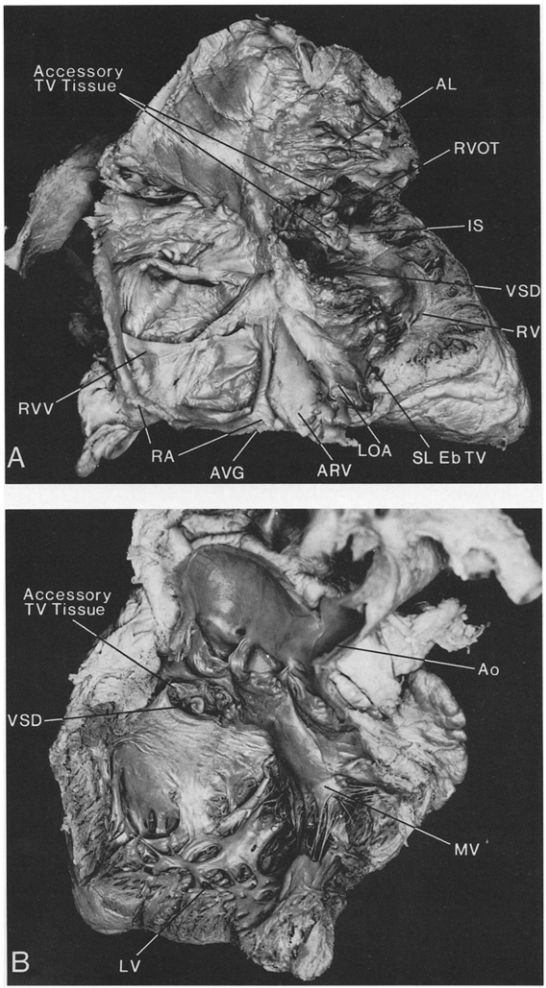

Figure 7. Case 5. A, View of the opened right atrium (RA) and right ventricle (RV). The right venous valve (RVV) of the inferior vena cava is unusually prominent. The atrialized portion of the right ventricle (ARV) can be seen between the atrioventricular groove (AVG) and the downwardly displaced septal leaflet of Ebstein's tricuspid valve (SL Eb TV). The anterior leaflet (AL) of the tricuspid valve is normally attached along the atrioventricular groove. The myxomatous accessory tricuspid valve (TV) tissue has been swung up and out of the ventricular septal defect (VSD). This defect is associated with anterior and superior displacement of the infundibular septum (IS) and narrowing of the right ventricular outflow tract (RVOT), typical of tetralogy of Fallot. B, View of the opened left ventricle (LV) and the mitral valve (MV), and of the opened aortic valve and proximal aorta (Ao). The ventricular septal defect (VSD) is almost occluded by the accessory tricuspid valve (TV) tissue associated with Ebstein's tricuspid valve.

deaths in our series, it is possible that ischemia was a contributing factor.

Conclusions. Echocardiography can accurately determine the presence and mechanism of obstructed ventricular septal defect in tetralogy of Fallot. In most cases, the septal defect is obstructed by accessory or excessive tricuspid valve tissue. This entity is important to recognize because of its apparent poor prognosis and suggests the need to modify the surgical technique.

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References

1. Van Praagh R, Van Praagh S, Nebesar RA, Muster AJ, Sinha SN, Paul MH. Tetralogy of Fallot: underdevelopment of the pulmonary infundibulum and its sequelae. Am J Cardiol 1970;26:25–33.
2. Rowe RD. Tetralogy of Fallot. In: Keith JD, Rowe RD, Vlad PJ, eds. Heart Disease in Infants and Children. New York: Macmillian, 1978:470–505.
3. Soulié P, Joly F, Carlotti J, Sicot JR. Etude comparee de l'hemodynamique dans les tetralogies et dans les trilogies de Fallot (etude de 43 cas). Arch Mal Coeur 1951;44:577–601.
4. McCord MC, Van Elk J, Blount SG Jr. Tetralogy of Fallot: clinical and hemodynamic spectrum of combined pulmonary stenosis and ventricular septal defect. Circulation 1957;16:736–49.
5. McGoon DC, Kirklin JW. Pulmonary stenosis with intact ventricular septum: treatment utilizing extracorporeal circulation. Circulation 1958;17:180–6.
6. Hoffman JIE, Rudolph AM, Nadas AS, Gross RE. Pulmonic stenosis, ventricular septal defect, and right ventricular pressure above systemic level. Circulation 1960;22:405–11.
7. Neufeld HN, McGoon DC, DuShane JW, Edwards JE. Tetralogy of Fallot with anomalous tricuspid valve simulating pulmonary stenosis with intact septum. Circulation 1960;22:1083–90.
8. Weis E, Fridman J, Shaffer AB. Tetralogy of Fallot with small ventricular septal defect. Acta Cardiol 1961;16:448–61.
9. Padmanabhan J, Varghese PJ, Lloyd S, Haller JA Jr. Tetralogy of Fallot with suprasystemic pressure in the right ventricle: a case report and review of the literature. Am Heart J 1971;805–11.
10. Van Praagh R, Ando M, Van Praagh S, et al. Pulmonary atresia: anatomic considerations. In: Kidd BSL, Rowe RD, eds. The Child with Congenital Heart Disease After Surgery. New York: Futura 1976:103–20.
11. Mesko ZG, Wagner HR, Subramanian S. Tetralogy of Fallot: occlusion of the ventricular septal defect due to accessory tricuspid valve leaflet and an associated membranous aneurysm. Eur J Cardiol 1978;7:257–62.
12. Fisher EA, Thanopoulos BD, Eckner FAO, Hastreiter AR, DuBrow IW. Pulmonary atresia with obstructed ventricular septal defect. Pediatr Cardiol 1980;1:209–17.
13. Faggian G, Frescura C, Thiene G, Borolotti U, Mazzucco A, Anderson RH. Accessory tricuspid valve tissue causing obstruction of the ventricular septal defect in tetralogy of Fallot. Br Heart J 1983;49:324–7.
14. LaCorte MA, Boxer RA, Singh S, Parnell V, Goldman M. Echocardiographic features of tetralogy of Fallot with an accessory tricuspid valve leaflet. Am Heart J 1985;110:1297–9.
15. Silver MD, Lam JHC, Rangnathan N, Wigle ED. Morphology of the human tricuspid valve. Circulation 1971;43:333–48.
16. Rosenquist GC, Sweeney LJ. Normal variations in tricuspid valve attachments to the membranous ventricular septum: a clue to the etiology of left ventricular to right atrial communication. Am Heart J 1975;89:186–8.
17. Davido A, Maarek M, Julien JL, Corone P. Maladie D'Ebstein associee a une tetralogie de Fallot: a propos d'une observation familiale, revue de la litterature, implication embryologique et genetique. Arch Mal Coeur 1985;78:752–6.
18. Chesler E, Korns ME, Edwards JE. Anomalies of the tricuspid valve, including pouches, resembling aneurysms of the membranous ventricular septum. Am J Cardiol 1968;21:661–8.
19. Varghese PJ, Izukawa T, Celermajer J, Simon A, Rowe RD. Aneurysm of the membranous ventricular septum: a method of spontaneous closure of small ventricular septal defect. Am J Cardiol 1969;24:531–6.
20. Tandon R, Edwards JE. Aneurysmlike formations in relation to the membranous ventricular septum. Circulation 1973;47:1089–97.
21. Anderson RH, Lenox CC, Zuberbuhler JR. Mechanisms of closure of perimembranous ventricular septal defect. Am J Cardiol 1983;52:341–5.
22. Ramaciotti C, Keren A, Silverman NH. Importance of (perimembranous) ventricular septal aneurysm in the natural history of isolated perimembranous ventricular septal defect. Am J Cardiol 1986;57:268–72.
23. Leung MP, Mok CK, Lo RNS, Lau KC. An echocardiographic study of perimembranous ventricular septal defect with left ventricular to right atrial shunting. Br Heart J 1986;55:45–52.
24. Deal BJ, Chin AJ, Sanders SP, Norwood WI, Castaneda AR. Subxiphoid two-dimensional echocardiographic identification of tricuspid valve abnormalities in transposition of the great arteries with ventricular septal defect. Am J Cardiol 1985;55:1146–51.
25. Chin AJ, Yeager SB, Sanders SP, et al. Accuracy of prospective two-dimensional echocardiographic evaluation of left ventricular outflow tract in complete transposition of the great arteries. Am J Cardiol 1985;55:759–64.
26. Gomes AS, Nath PH, Singh A, et al. Accessory flaplike tissue causing ventricular outflow obstruction. J Thorac Cardiovasc Surg 1980;80:211–6.
27. Van Praagh R, Corwin RD, Dahlquist EH Jr, Freedom RM, Mattioli L, Nebesar RA. Tetralogy of Fallot with severe left ventricular outflow tract obstruction due to anomalous attachment of the mitral valve to the ventricular septum. Am J Cardiol 1970;26:93–101.
28. Van Praagh R, Van Praagh S. The anatomy of common aorticopulmonary trunk (truncus arteriosus communis) and its embryologic implications: a study of 57 necropsy cases. Am J Cardiol 1965;65:406–25.
29. Rosenquist GC, Sweeney LJ, Stemple DR, Christianson SD, Rowe RD. Ventricular septal defect in tetralogy of Fallot. Am J Cardiol 1973;31:749–54.
30. Goor DA, Lillehei CW. Dextroposition of the aorta. In: Goor DA, Lillehei CW, eds. Congenital Malformations of the Heart. New York: Grune & Stratton, 1975:169–79.
31. Freedom RM, White RD, Pieroni DR, Varghese PJ, Krovetz LJ, Rowe RD. The natural history of the so-called aneurysm of the membranous ventricular septum in childhood. Circulation 1974;49:375–84.
32. Hoeffel JC, Henry M, Flizot M, Lucieri R, Pernot C. Radiologic patterns of aneurysms of the membranous septum. Am Heart J 1976;91:450–6.
33. Burrows PE, Fellows KE, Keane JF. Cineangiography of the perimembranous ventricular septal defect with left ventricular-right atrial shunt. J Am Coll Cardiol 1983;1:1129–34.
34. Mirsky I, Laks MM. Time course of changes in the mechanical properties of canine right and left ventricles during hypertrophy caused by pressure overload. Circ Res 1980;46:530–42.
35. Wessel HU, Weiner MD, Paul MH, Bastanier CK. Lung function in tetralogy of Fallot after intracardiac repair. J Thorac Cardiovasc Surg 1981;82:616–28.
36. Bove EL, Kavey RW, Byrum CJ, Sondheimer HM, Blackman MS, Thomas FD. Improved right ventricular function following late pulmonary valve replacement for residual pulmonary insufficiency or stenosis. J Thorac Cardiovasc Surg 1985;90:50–5.
37. Sauer V, Bindl L, Pilossoff V, Hultzsch W, et al. Pulmonary atresia with intact ventricular septum and right ventricule-coronary artery fistulae:

selection of patients for surgery. In: Doyle EF, Engle MA, Gersony WM, Rashkind WJ, Talner NS, eds. Pediatric Cardiology Proceedings of the Second World Congress of Cardiology. New York: Springer-Verlag, 1986:566–78.

38. Fyfe DA, Edwards WD, Driscoll DJ. Myocardial ischemia in patients with pulmonary atresia and intact ventricular septum. J Am Coll Cardiol 1986;8:402–6.