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 entity (2-14) with a relatively high operative mortality rate (6-9). 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 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 7020 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.

Direct observation 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 the same year.

Results

Incidence and clinical course (Table I). Between January 1982 and February 1986, 269 patients with unrepaired tetral-
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 wholly 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 61% (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 arrow) can be seen separately in the right ventricle (RV). Ao = aorta; i = inferior; l = left; r = right; s = superior.

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 valvulike 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 annulus 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 Chester 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 valvar aortic stenosis.

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 reported 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 arrhythmia 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
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
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