the development of the cardiac and pulmonary vascular complications which were observed in these three children. Nine to thirteen years ago when the original mitral valve prostheses were introduced, only the Starr-Edward ball-cage valve was available. Since that time a number of other artificial valves have been offered for clinical use. In view of the development of left ventricular wall thickening in all three cases and the extension of fibrous tissue onto the valve struts in one, it would seem wise to avoid the use of prosthetic valves which may partially occlude the left ventricular outflow tract in small children. The glutaraldehyde-preserved porcine heterograft currently possesses the greatest number of advantages for use in this age group because of its central flow characteristics, favorable effective orifice to anular diameter ratio, low thromboembolic rate without anticoagulants, and proven durability of five or more years. References


SUMMARY Absent pulmonic valve (APV) in tetralogy of Fallot produces a pulmonic regurgitation murmur (PRM) which is usually late in onset after A2, low pitched, and of crescendo-decrescendo character. We have seen three adult patients with tetralogy of Fallot with APV and have done intracardiac sound and pressure studies in two. The PRM was loudest in the RV outflow tract (RVOT), where the onset was earlier than the murmur recorded on the chest wall. The crescendo portion of the PRM occurred during an abnormally slow decline in the RVOT pressure pulse after the crossover of PA and RVOT pressures. The RVOT pressure reached its minimum 30 msec after the RV body pressure, resulting in a pressure gradient between the two. The PRM peaked 30 msec later in the RV body than in the RVOT. The delayed precordial onset of the PRM after A2, is likely due to failure of transmission of early vibrations through the chest wall. The morphology of the PRM in tetralogy of Fallot with APV may be related to delayed relaxation with altered diastolic compliance of the RVOT which is subjected to a large regurgitant volume from the massively dilated pulmonary arteries.

TETRALOGY OF FALLOT with absent pulmonic valve is an entity which can be differentiated from the classical tetralogy of Fallot. Distinctive features are those of right ventricular dilatation as well as hypertrophy, obstruction usually at a small pulmonic valve ring with absent or mild infundibular obstruction, absent pulmonic valve cusps, and aneurysmal proximal pulmonary arteries. On clinical examination the harsh systolic ejection murmur along the left sternal border and absent pulmonic second sound generally occur in classic tetralogy, but the low pitched, crescendo-decrescendo murmur of pulmonic regurgitation heard along the left sternal border is not heard in unoperated tetralogy of Fallot.

The determinants of the characteristics of the pulmonic regurgitation murmur in tetralogy of Fallot with absent pulmonic valve have not been documented.

Three adults in whom the diagnosis was established by operation and/or autopsy had complete clinical, phonocardiographic, and catheterization studies. In addition, two had intracardiac sound and pressure studies performed to define the mechanism of production of the murmur.

Materials and Methods

Diagnostic cardiac catheterization was performed with standard fluid-filled catheter systems, using hydrogen.

References


platinum electrode catheters for shunt detection, and oximetry and nitrous oxide inhalation for shunt quantitation. Meglumine and sodium diatrizoate was used for all angiograms. External phonocardiograms were recorded on a Sanborn 550 recorder using Sanborn contact crystal microphones and 350-1700B heart sound preamplifiers. Intracardiac sound and pressure recordings were obtained in cases 2 and 3 using a Telec micromanometer tipped catheter. Recordings were made on a Sanborn 550M or a Hewlett Packard 4568C recorder using bioelectric DC amplifiers. The frequency response of the system was flat to 200 Hz. All recordings are shown at a paper speed of 100 mm/sec.

**Results**

The phonocardiograms of the three patients are shown in figure 1. They illustrate the classic findings in patients with tetralogy of Fallot with absent pulmonic valve. The prominent systolic ejection murmur extending into the aortic second sound was recorded best along the left sternal border. A quiet interval of 50 to 100 msec was followed by a low pitched crescendo-decrescendo diastolic murmur best recorded along the upper and mid-left sternal border. No pulmonic component of the second sound could be identified.

The catheterization (table 1) and angiographic findings were also typical of the entity: a pressure gradient predominately at the level of the pulmonic valve anulus, left-to-right shunting without significant right-to-left shunting through the ventricular septal defect, low pulmonary artery diastolic pressure consistent with pulmonic regurgitation, right ventricular hypertrophy and dilatation, and aneurysm dilatation of the pulmonary arteries.

Analysis of the characteristics of the pulmonic regurgitation murmur in relation to the pressure pulses obtained from the pulmonary artery, right ventricular outflow tract, and right ventricular inflow tract resulted in a reasonable explanation of the determinants of the murmur characteristics.

Figure 2 shows the intracardiac phonocardiograms from patients 2 and 3 recorded in the right ventricular outflow tract. The crescendo-decrescendo murmur was clearly earlier in onset within the right ventricular outflow tract than was discerned by either auscultation or external phonocardiography most likely due to failure of transmission of the early low frequency vibrations through the chest wall.

Superimposition of the manometer pressure pulse recordings of identical cycle lengths from the pulmonary artery and right ventricular outflow tract (fig. 3) demonstrated that in both cases the onset of the murmur occurred with the development of a pressure gradient between the pulmonary artery and RV outflow tract. The murmur reached peak intensity when the pulmonary artery-RV outflow tract gradient was maximum and then gradually

**Table 1. Cardiac Catheterization Data**

<table>
<thead>
<tr>
<th></th>
<th>RA</th>
<th>RVIT</th>
<th>RVOT</th>
<th>MPA</th>
<th>SA</th>
<th>L → R shunt</th>
<th>SA</th>
<th>O₂ sat.</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>S.M.</td>
<td>(1)</td>
<td>97/0</td>
<td>90/0</td>
<td>35/0(16)</td>
<td>101/61</td>
<td>1.5:1</td>
<td>86%</td>
</tr>
<tr>
<td>2</td>
<td>D.R.</td>
<td>(8)</td>
<td>93/8</td>
<td>25/4(9)</td>
<td>100/60</td>
<td>1.5:1</td>
<td>96%</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>T.C.W.</td>
<td>(3)</td>
<td>96/6</td>
<td>76/6</td>
<td>20/8</td>
<td>98/56</td>
<td>2:1</td>
<td>93%</td>
</tr>
</tbody>
</table>

Abbreviations: RA = right atrium; RVIT = right ventricular inflow tract; RVOT = right ventricular outflow tract; MPA = main pulmonary artery; SA = systemic artery; L → R = left to right.
diminished in intensity as the gradient diminished, terminating at the point of equilibration of the pressures.

The right ventricular outflow tract pressure pulse showed an abnormally slow decline in pressure after the crossover of the pulmonary artery and RV outflow tract pressures (fig. 4). This slow pressure decline was responsible for the slow development of the maximum pulmonary artery-RV outflow tract diastolic gradient, which in turn corresponded to the crescendo portion of the murmur. The RV outflow tract pressure pulse differed from the RV inflow tract pressure pulse as can be seen in figure 4, which compares pressures from the two sites during equal cycle lengths. In both cases the outflow tract pressure took 30 msec longer to reach minimum diastolic pressure than the inflow tract pressure, creating an early diastolic pressure gradient between the outflow tract and the RV inflow tract, the outflow tract pressure being higher.

In patient 3, recordings of the murmur from the outflow tract demonstrated the earlier onset of the pulmonic regurgitation (DM) internally compared to externally. 4 LSB = 4th left intercostal space, F-200 = frequency cut-off 200 Hz, RVOT = right ventricular outflow tract, SM = systolic murmur, DM = diastolic murmur.

**Figure 2.** External (top) and intracardiac (IC) phonocardiograms from cases 2 (left) and 3 (right) from the right ventricle demonstrating the earlier onset of the pulmonic regurgitation (DM) internally compared to externally. 4 LSB = 4th left intercostal space, F-200 = frequency cut-off 200 Hz, RVOT = right ventricular outflow tract, SM = systolic murmur, DM = diastolic murmur.

**Figure 3.** Intracardiac phonocardiograms and pressure pulses from the right ventricular outflow tract (RVOT) from cases 2 (left panel) and 3 (right panel) with superimposed manometer pressure pulses from the pulmonary artery (PA) demonstrating the relationship between the murmur configuration and the PA-RVOT gradient.
tract and from the body of the right ventricle below the infundibulum demonstrated that the murmur peaked 30 msec later in the body of the right ventricle than in the outflow tract. Satisfactory recordings just below the outflow tract were not obtained in patient 2.

Discussion

The relationship between the murmur configuration and the appearance and disappearance of a pulmonary artery-right ventricular outflow tract diastolic gradient was also noted by Harris et al. in 1962 in a 22-year-old patient with tetralogy of Fallot with absent pulmonic valve. Fluid-filled catheters were used, so further pressure pulse characteristics could not be defined. Bousvaros and Deuchar did not discuss pressure pulses in their intracardiac sound studies in patients with pulmonic regurgitation, one of whom may have had an absent pulmonic valve. Intracardiac sound and pressure studies performed with manometer catheters have been reported in isolated pulmonic regurgitation due to bacterial endocarditis, one of which demonstrated a gradient-murmur relationship similar to that seen in our patients. The pressure pulse morphology in the pulmonary artery and right ventricle differs considerably between the postendocarditis patients and ours, however.

In our patients with tetralogy of Fallot with absent pulmonic valve, the diastolic pressure gradient between the pulmonary artery and right ventricular outflow tract is very small, because of the very low pulmonary pressure due to valve ring stenosis. The crescendo portion of the pulmonic regurgitation murmur has considerable duration, however, because of the delayed fall in right ventricular outflow tract pressure after the crossover with the pulmonary artery pressure. In patients with isolated pulmonary valve regurgitation, the pulmonary artery pressure is higher and the right ventricular pressure then falls considerably farther to minimum diastolic pressure. The crescendo portion of the murmur occurs during this greater and more rapid decrease in right ventricular pressure. The delayed fall in early diastolic pressure in the right ventricular outflow tract in our patients appears to be unique. We have not seen similar pressure pulse morphology in patients with classical tetralogy of Fallot, isolated valvular pulmonic stenosis before or after operation, idiopathic dilation of the pulmonary artery, or pulmonic regurgitation due to severe pulmonary hypertension who have been studied with manometer tipped catheters in our laboratory.

The delayed fall in early diastolic pressure in the right ventricular outflow tract may be explained by increased diastolic stiffness and impaired relaxation of the musculature of the infundibulum, most particularly the crista supraventricularis, which has been noted to be hypertrophied in pathological studies reported in patients with tetralogy of Fallot with absent pulmonic valve. Reduced compliance of muscle secondary to hypertrophy and fibrosis would likely delay relaxation even more. In these patients, then, the blood regurgitating into the outflow tract in diastole due to absence of the pulmonic valve may be temporarily retained or slowed in its course back into the body of the right ventricle, which results in a decrease in the rate of pressure fall in the outflow tract and prolongation of the time to reach minimum diastolic pressure. An actual diastolic pressure gradient exists between the right ventricular outflow tract and inflow tract as a result. The probability of obstruction to regurgitant flow from infundibulum to body of the right ventricle in these patients is further substantiated by the finding of a delay in the time to reach peak murmur amplitude in the right ventricular body compared to the outflow tract in one of our patients (case 3).

Other possible contributing factors to the delay in transmission of the regurgitant volume from outflow tract to inflow tract could be 1) angulation between the infundibulum and body of the right ventricle seen in some children with the lesion under discussion, but not present in our patients and 2) a right intraventricular conduction defect, which was present in only one of our patients, although both had similar pressure pulses.

The question could be raised that the determinants of the murmur characteristics as described here may not apply to other patients with tetralogy of Fallot with absent pulmonic valve because many of the descriptions of the pulmonary valve regurgitation murmur are of "descrescendo" murmurs or are described as the "fro" of a to and fro murmur. The crescendo portion of the murmur may not be recognized due to failure of transmission of the low frequency components to the chest wall as demonstrated in our patients. In addition, since most reported cases are in infants and children, the crescendo portion may not be recognized due to rapid heart rates and extraneous noise.

Our conclusions concerning the timing and configuration of the murmur of pulmonic regurgitation in adult patients

Figure 4. Comparisons of the manometer pressure pulses from the right ventricular outflow tracts (RVOT) (top panels) and right ventricular inflow tracts (RVIT) (bottom panels) from equal cycle lengths in cases 2 (left) and 3 (right). Note the delay in reaching minimum pressure in the RVOT compared to RVIT in both cases.
with tetralogy of Fallot with absent pulmonic valve are as follows:

1) The murmur of pulmonic regurgitation is of earlier onset on intracardiac recordings than can be heard externally due to failure of transmission of the initial low frequency vibrations to the chest wall.

2) The crescendo-decrescendo murmur appears to be related to the development of and the disappearance of a pulmonary artery-right ventricular outflow tract gradient in diastole.

3) The development of the pulmonary artery-right ventricular outflow tract gradient is primarily related to an abnormally slow decline in right ventricular outflow tract pressure.

4) The abnormally slow decline in right ventricular outflow tract pressure, which also results in an early diastolic pressure gradient between the outflow tract and the right ventricular inflow tract, is most likely related to hypertrophy with accompanying increased stiffness and/or delayed relaxation of the infundibular chamber causing temporary retention of the regurgitant volume.

References


10. Kurtz CM, Sprague HB, White PD, Congenital heart disease, interventricular septal defects with associated anomalies in a series of 3 cases examined post mortem, and a living patient 58 years old with cyanosis and clubbing of the fingers. Am Heart J 3: 77, 1927


The murmur of pulmonic regurgitation in tetralogy of Fallot with absent pulmonic valve.
M E Fontana and C F Wooley

Circulation. 1978;57:986-990
doi: 10.1161/01.CIR.57.5.986

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
Copyright © 1978 American Heart Association, Inc. All rights reserved.
Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/57/5/986