CLINICOPATHOLOGIC CORRELATIONS

Aortopulmonary Septal Defect Coexisting with Ventricular Septal Defect

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SUMMARY

Three cases, in each of which an aortopulmonary septal defect (AP window) coexisted with a ventricular septal defect, are described. In two the ventricular septal defect was of the usual variety, and in one it was part of the tetralogy of Fallot.

The patients presented as examples of congestive cardiac failure with a large left-to-right shunt in infancy. Aortography was the most reliable method for identification of the AP window when a ventricular septal defect was associated. Associated features included a right aortic arch with mirror image branching in the first case and tetralogy of Fallot with muscular subaortic stenosis in the third case. In the case with the tetralogy of Fallot, the effects of pulmonary stenosis were masked by the left-to-right shunt through the AP window.

Additional Indexing Words:
Left-to-right shunt
Tetralogy of Fallot
Muscular subaortic stenosis
Right aortic arch

Among patients with clinically evident left-to-right shunts occurring either between the ventricles or between the great arteries, the prevailing conditions are isolated ventricular septal defect, accounting for the former, and patent ductus arteriosus, for the latter. The tendency for these two conditions to dominate the state of a left-to-right shunt is so strong that usually, on the basis of chance alone, the ultimate diagnosis of either is correct. Yet, more complicated situations may be encountered. For example, among subjects with ventricular septal defect without coarctation of the aorta, Girod and associates found an associated patent ductus arteriosus in about 9 per cent of the cases.

Moreover, when either ventricular septal defect or patent ductus initially is considered to be the basis for a left-to-right shunt, the fundamental condition may be more complex. For example such conditions as persistent truncus arteriosus, single ventricle or double outlet right ventricle clinically may simulate isolated ventricular septal defect and aortopulmonary septal defect (AP window) may simulate patent ductus arteriosus. Further complexities may occur when a given condition which usually occurs as an isolated entity is associated with yet another condition that also allows a left-to-right shunt.

In the latter regard, AP window is a condition in point. This condition is characterized as a window-like communication between the ascending aorta and pulmonary trunk. Although no other defects are present in the classical case, there are exceptions. The purpose of this communication is to emphasize these. Among 66 cases of AP window reviewed from the literature by Neufeld and associates, there were eight examples in which a patent ductus arteriosus was associated, this state being the most common association found in that series.

With regard to the association of ventricular septal defect with AP window, isolated cases have been reported. We have observed three such cases, in one of which the ventricular septal defect was part of the tetralogy. In this case, the left-to-right shunt through the AP window served to modify the effects of the tetralogy of Fallot.

We shall review the clinical and pathologic features.
of our three cases of AP window associated with ventricular septal defect, one of which was the case with the tetralogy of Fallot mentioned.

**Cases with Ventricular Septal Defect**

**Clinical Features**

The first case was an 18-month-old girl with cardiomegaly and a history of respiratory infection in early infancy. Physical examination revealed a grade IV/V, pansystolic murmur along the left sternal border and an accentuated pulmonary component of the second cardiac sound. Thoracic roentgenograms showed cardiomegaly with increased pulmonary vasculature and a right aortic arch. Biventricular hypertrophy was evident in the electrocardiogram. Cardiac catheterization and angiograms indicated the presence of a large left-to-right shunt through a ventricular septal defect. An aortogram was read as showing the left pulmonary artery arising from the ascending aorta.

The preoperative diagnosis was ventricular septal defect and origin of the left pulmonary artery from the ascending aorta. The operation, which was done without extracorporeal circulation, had been planned to anastomose the left pulmonary artery to the pulmonary trunk and to band the latter. During the operation it was found that the pulmonary arteries arose normally, while an AP window was present. This was closed without either of the great vessels being opened and the pulmonary artery was banded. Death occurred shortly after completion of the procedure.

The second case (case 2) involved an 8-year-old girl in whom a murmur was noted at the age of six weeks when she was examined because of failure to thrive. At that time, physical examination suggested a large left-to-right shunt at the ventricular level and congestive cardiac failure. Cardiomegaly and increased pulmonary vasculature were evident in the thoracic roentgenograms. In the electrocardiogram, the mean QRS axis was +105° with biventricular hypertrophy. Cardiac catheterization studies indicated a left-to-right shunt at the ventricular level. The patient was not seen again until the age of three years when she appeared well. At this time, there was a loud pansystolic murmur and accentuated pulmonary component of the second sound. The patient was restudied because of pulmonary arterial hypertension. The systolic pressure in both ventricles and both great vessels was identical. At the ventricular level, a left-to-right shunt with the pulmonary flow more than twice the systemic flow was found.

A right ventriculogram opacified the base of the left ventricle and both great vessels (fig. 1). An AP window was identified in an aortogram. The patient was re-evaluated at the age of eight years, at which time she was still asymptomatic and the physical findings were unchanged. At this time, studies showed the pulmonary and systemic arterial pressures to be identical at 105/70 mm Hg. The pulmonary flow was 7.8 L/min and the systemic flow 3.1 L/min. There was no right-to-left shunt. The ventricular septal defect and the AP window were closed surgically. Death occurred in the early postoperative period.

**Pathologic Features**

In both cases 1 and 2, in addition to evidence of the operative procedures described, there was a ventricular septal defect of the usual variety and classical example of AP window (figs. 2 and 3). Additional details were found in case 1. There was a right aortic arch with mirror image branching and pouch-like accessory tissue involving the septal leaflet of the tricuspid valve (fig. 2).

**Comment**

In each case, the clinical picture and the findings at cardiac catheterization were compatible with a left-to-right shunt occurring through an isolated ventricular septal defect. In case 2 the AP window was identified only by aortography. In case 1, while an aortogram had been performed, it was misinterpreted as origin of the left pulmonary artery from the aorta.

Regardless of the latter error in diagnosis, aortography remains as the reliable method of identifying an AP window beyond a ventricular septal defect. Studies of oxygen saturation generally cannot be expected to identify the AP window when a ventricular septal defect lies proximal to it; when two sites of a left-to-right shunt occur, the proximal one is identifiable while the increased level of oxygen saturation
Case 1. a. Interior of the right ventricle (R.V.). The ventricular septal defect (probe) lies postero-inferior to the parietal band of the crista supraventricularis (C.S.) and beneath the septal leaflet of the tricuspid valve (S.T.). P.V. = pulmonary valve. b. Interior of the left ventricle (L.V.) and aorta (A.). The ventricular septal defect (D.) occupies the area of membranous septum and part of the anterior smooth septum. c. Great vessels from in front. Sites of operative intervention (between arrows) represent end result of closure of AP window as seen in the pulmonary trunk (P.T.). A. = aorta.

it causes obscures signs of the second, downstream shunt.

The presence of a right aortic arch in our first case is uncommon as a condition associated with AP window. Neufeld and associates2 found two examples among 66 cases of AP window.

Case with Tetralogy of Fallot

Clinical Features

The third case (case 3)* was that of a 3-month-old male infant who was referred because of fatigability while eating and excessive perspiration. Physical examination showed an acyanotic child in a state of congestive cardiac failure. The heart was enlarged. A grade III/VI, pansystolic murmur was present along the left sternal border. An apical diastolic flow murmur was heard. The pulmonary component of the second cardiac sound was accentuated. The electrocardiogram showed a mean QRS axis of +90° with biventricular hypertrophy. A thoracic roentgenogram revealed cardiomegaly and increased pulmonary vasculature. Cardiac catheterization showed the following pressures: left ventricle, 120/0-13; aorta, 70/40; right ventricle, 70/0. Oxygen saturations in

*Details of this case are being reported elsewhere (Marin-Garcia and associates: Am Heart J, in press).

Figure 2

Case 2. a. Interior of the right ventricle (R.V.). The ventricular septal defect (D.) lies postero-inferior to the crista (C.S.). P.V. = pulmonary valve. b. Interior of the left ventricle (L.V.) and ascending aorta (A.). The ventricular septal defect (D.) occupies the area of membranous septum. The suture line (between arrows) in the aorta indicates the site of the surgically closed AP window.

percentages were as follows: left atrium, 96; aorta, 88; right ventricle, 36.

The catheter was not advanced into the pulmonary trunk. During the same study, a right ventriculogram was performed. This showed subpulmonary stenosis and a right-to-left shunt into the aorta with an unusual degree of opacification of the pulmonary arteries, in view of the presence of pulmonary stenosis. The left anterior oblique view of this study showed a communication between the ascending aorta and the pulmonary trunk (fig. 4). The latter served to establish the presence of an AP window. Aortography also confirmed the presence of an AP window.

Figure 3

Figure 4

Case 3. Right ventriculogram. a. Right anterior oblique view. Simultaneous opacification of the aorta (A.) and the narrow right ventricular outflow tract (arrow). b. Left anterior oblique view. The AP window (between arrows) lies proximal to the branches of the aortic arch. R.V. = right ventricle.

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Shortly after the studies, the infant developed cardiopulmonary arrest and died.

Pathologic Features

The necropsy showed the usual features of the tetralogy of Fallot but with an unusually wide pulmonary trunk. Study of the interior of the great vessels showed a classical example of AP window. Also, the ventricular septum showed marked asymmetrical hypertrophy resulting in severe subaortic stenosis (fig. 5).

Comment

In this case with the tetralogy of Fallot, the left-to-right shunt through the associated AP window served to alter significantly the clinical picture usually seen in the tetralogy. Clinically, the dominant condition appeared to be one of a left-to-right shunt. While both aortography and right ventriculography identified the AP window, the latter study showed the unexpected finding of the presence of the tetralogy of Fallot. This case also showed hypertrophic muscular subaortic stenosis, a condition which is uncommon in association with either the tetralogy of Fallot or AP window.

To our knowledge, there are two reported instances of subaortic stenosis associated with AP window. In one of these, subaortic stenosis was observed clinically and its anatomic type is unknown, while in the other (Somerville’s case 5) the membranous type was identified at necropsy.

We are aware of only one other reported case of AP window in association with the tetralogy.

General Comment

AP window is an uncommon malformation which classically is not associated with other significant defects. While this is true for the majority of cases, there are exceptions. Patent ductus arteriosus has been identified in about 12 per cent of cases with AP window. Ventricular septal defect is rarely associated.

When ventricular septal defect and AP window are associated, oxygen studies tend, as in our cases 1 and 2, to identify only the ventricular septal defect. The ideal method of identifying an associated AP window appears to be aortography.

The presence of an AP window in association with ventricular septal defect does not alter the basic hemodynamics of ventricular septal defect.

In contrast, as in our case 3, the left-to-right shunt of an AP window associated with the tetralogy of Fallot tends to obscure the classical features of the tetralogy.

References

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