Congenital Pulmonary Stenosis Resulting from Dysplasia of Valve

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Colin J. Schwartz, M.D., and Jesse E. Edwards, M.D.

SUMMARY
A distinctive type of pulmonary valvular stenosis, termed “pulmonary valvular dysplasia,” is described from six necropsies and 10 living children. The anatomic features of the stenotic pulmonary valve are unique in that there are three distinct cusps and no commissural fusion. The obstructive mechanism is related to markedly thickened, immobile cusps, characterized by the presence of disorganized myxomatous tissue.

Several clinical features tend to distinguish this form of pulmonary stenosis. In each of 16 patients studied, a pulmonary ejection murmur was present, but this was not associated with an ejection click. Other features suggestive of this type of pulmonary stenosis were slow body growth, abnormal facies, and a positive family history of pulmonary stenosis. The electrocardiogram showed a greater degree of right axis deviation than is found in most cases of dome-shaped pulmonary stenosis. Right ventriculography revealed distinctive features of the pulmonary valve which were characterized by lack of typical dome-shaped deformity and by the presence of thick cusps.

Experience with simple incision-valvulotomy was associated with a high (38%) operative mortality and significant residual stenosis in survivors. The suggested operative procedures include excision of a valve cusp, placement of a right ventricular outflow patch, or replacement of the pulmonary valve.

Additional Indexing Words:
Pulmonary valvular stenosis Electrocardiography in pulmonary stenosis

Among patients with congenital pulmonary valvular stenosis, there are isolated instances in which the nature of the pulmonary valve differs from the well-recognized dome-shaped deformity.

Among the exceptions is a deformity characterized by the presence of three distinct cusps and commissures, but in which the valvular tissue is abnormally thick, the principal feature in causing the valve to be stenotic. We have applied the term “pulmonary valvular dysplasia” to this entity.

It is the purpose of this communication to describe the pathologic features in six cases studied with necropsy and to relate this experience to the clinical findings. As this material served to make us aware of this entity, we identified pulmonary valvular dysplasia in 10 additional patients, each of whom is living. The clinical experience of the 16 patients (including the six studied at necropsy) will be described to indicate the features which serve to distinguish patients with pulmonary valvular dysplasia and pulmonary stenosis from those with classic dome-shaped pulmonary valvular stenosis.

Observations
In order that the clinical manifestations of pulmonary valvular dysplasia be most mean-

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Table 1
Summary of Cardiac Lesions Associated with Pulmonary valvular dysplasia in Six Cases Studied at Necropsy.

<table>
<thead>
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<th>Case</th>
<th>Atrial Septal Defect</th>
<th>Infundibular pulmonary stenosis</th>
<th>Supravalvular pulmonary stenosis</th>
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* Thickened aorta
** Ventricular septal defect
Present
Absent

In each of the six patients who died, the ages of whom ranged from 4 months to 17 years, the pulmonary valve was essentially similar. Five of these patients died after attempted surgical relief of pulmonary stenosis, while the sixth patient (case 1) died suddenly at home. At necropsy in each case, the pulmonary valve showed three distinct leaflets and comissures. As viewed from above, the gray-white cusps were greatly thickened, shortened, and rigid (fig. 1).

Additionally, in most of the cases, a prominent, padlike fibrous mass occupied the depths of a sinus of the valve (fig. 1b). Such masses prevented lateral excursion of the related cusp.

In the interest of explaining the histologic characteristics of the alterations in the cusps, reference to the normal is appropriate.

In the normal pulmonary valve, each cusp shows an intermediate layer of loose connective tissue termed the "spongiosa" (fig. 2a). This layer is separated from the ventricular

Figure 1
(a) Case 4. Pulmonary valve viewed from above. Three distinct cusps. Each thicker than usual.
(b) Case 3. Horizontal section near inferior level of valve. Each of three cusps greatly thickened.
A padlike thickening (P.) of tissue lies in a sinus.
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(a) Low-power photomicrograph of a normal pulmonary valve. V = ventricularis; S = spongiosa. Movat stain; × 67. (b) Case 3. Photomicrograph of pulmonary valve at level comparable to horizontal section depicted in figure 1b. Thickening of cusps is apparent. There is excessive, pale, loose connective tissue in the spongiosa as well as beneath the arterial surface of the leaflet. In the padlike thickening (P.), one sinus is adherent to the related cusp. Elastic tissue stain; × 6.

and arterial surfaces of the cusp by more dense fibrous layers termed the "ventricularis" and "fibrosa," respectively. In the specimens under consideration the essential change was major thickening of the spongiosa, a process which, for the most part, accounted for thickening of the individual cusps noted grossly (fig. 2b). The tissue which caused the spongiosa to be thickened (fig. 3) was composed of stellate and fusiform cells lying in a basophilic matrix, the picture resembling embryonic connective tissue. In some instances, capillaries and larger vessels were present in the thickened tissue and, in foci, the abnormal tissue of the spongiosa encroached upon and interrupted the other layers, especially the ventricularis. The pads of tissue which occupied some of the deepest parts of the valve sinuses were continuous with the related thickened cusp and composed of the same type of abnormal tissue as in the cusp.

In one case, the histologic changes in the cusps were not uniform. Some areas showed thickening of the spongiosa and fibrosa as described while, in intervening areas, the structure was normal. Uninvolved zones were seen in several areas and tended to occur toward the base of the cusp.

In one case (case 1) coincidental involvement of the aortic valve was observed. The histologic appearance of the aortic valve was similar to that observed in the pulmonary valve. In this case, the walls of the pulmonary trunk and aorta were grossly thickened. Histologically, the great arterial vessels revealed medial hypertrophy, loss of the normal parallel orientation of the elastic laminae, and replacement by an interlacing mosaic of medial elements. This abnormality resembled the changes seen in supravalvular aortic stenosis.
although no distinct obstruction of the great vessels was apparent.

In another case (case 3) there was stenosis at the bifurcation of the pulmonary trunk, as well as stenosis at the origin of several branches of the right and the left pulmonary arteries (fig. 4). Histologically, the lesions were represented by intimal fibrous thickening without alteration of the underlying media. Beyond foci of stenosis, the involved vessels were dilated.

In each of the six cases, prominent right ventricular hypertrophy was present, and three patients exhibited infundibular stenosis as a result of muscular hypertrophy.

In three of the six cases, a valvular competent patent foramen ovale was present, while in each of the remaining three, a true defect was observed in the atrial septum. In one of the latter, a ventricular septal defect was also present. Two of the atrial septal defects were of the fossa ovalis type, while in the third case (no. 6), the defect was located in the lowermost part of the atrial septum. In the latter case, an accessory orifice was present in the mitral valve.
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Figure 4

Case 3. Peripheral pulmonary arterial stenosis. (a) Interior of opened pulmonary trunk and right (R. P. A.) pulmonary artery. A circumferential rim of intimal thickening (between arrows) represents an obstruction at the origin of the right pulmonary artery. The ostium of the left pulmonary artery (probe below L. P. A.) is greatly narrowed. (b) Peripheral intrapulmonary arteries. At two points (each between paired arrows) circumferential intimal thickening causes stenosis of the ostium of the involved branch.

Clinical Features (Table 2)

The clinical and laboratory features of the six patients studied at necropsy and of the 10 living patients will be described together. In the 10 living patients the diagnosis of pulmonary stenosis related to pulmonary valvular dysplasia was established on the basis of the clinical findings and the characteristic angiographic features of the pulmonary valve.

Each of the patients presented a loud pulmonary systolic ejection murmur characteristic of pulmonary valvular stenosis and, in addition, four other features tended to recur. These were (1) retarded body growth, (2) absence of a pulmonary systolic ejection click, (3) abnormal facies, and (4) a positive family history for pulmonary stenosis.

Among the 16 patients, 10 were male and six were female. In the necropsy group, the ages at death ranged from 4 months to 17 years (table 2). In four of these six, the ages were 6½ years or older, the next youngest patient being 2½ years old. At present, the living patients range in age from 6 to 14 years. The latter patients have been observed for periods varying from 2 to 11 years.

Although none of the 16 patients presented signs of congestive cardiac failure, shortness of breath, or easy fatigability was described

Table 2

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* In no case was a pulmonary ejection click noted.
in nine instances. In five of these nine, these symptoms were evident during infancy and were especially notable during feeding. Cyanosis was present in only three patients, and in each it was mild (cases 10, 12, and 16). Cyanosis was first evident during infancy in two of these three patients and appeared at the age of 3 years in the third patient (case 16).

The pulmonary murmur present in each patient usually constituted the basis for suspecting congenital heart disease. In 10 patients, the murmur was noted in the first month of life. In the remaining patients, it was heard initially at the ages of 5 months, 6 months, 1 year, 2 years (two cases) and 3 years, respectively. The pulmonary murmur was associated with a thrill in 14 patients. In no patient was a systolic ejection click or diastolic murmur evident. The pulmonic component of the second sound was soft or absent in each patient.

A family history of pulmonary stenosis was present in four patients (cases 2, 9, 11, and 16). In case 2, the mother and a sister presented findings of pulmonary valvular stenosis. The mother, who is small of stature and has an abnormal facies, was demonstrated at operation to have a typical domed type of pulmonary stenosis. The details of the valve are unknown to us in the case of the sister.

A brother of patient 9 exhibited typical pulmonary valvular dome-shaped stenosis, requiring pulmonary valvulotomy. A sister of patient 16 presented clinical and catheterization evidence of mild pulmonary valvular stenosis. In neither of the latter cases do the involved siblings present the clinical or laboratory features considered typical of pulmonary valvular dysplasia. Patient 11 was a member of a family in which the mother and a male sibling present evidence of mild pulmonary stenosis and a markedly abnormal electrocardiographic QRS axis.

In the three other families of our patients, the histories revealed two miscarriages in one family, three in a second, and two miscarriages and two stillbirths in the third. In a single instance (case 3), a history of maternal mumps during the first trimester of pregnancy was given.

Fourteen of the 16 patients exhibited retardation of growth despite the facts that congestive cardiac failure had not been present in any case and that only four were premature according to body weight.

Thirteen of the patients were below the tenth percentile for height and weight and one at the sixteenth percentile. The body size of the remaining two patients was normal.

An abnormal facial appearance was observed in eight of the patients, five of whom were male and three female. The face was triangular in shape, with hypertelorism, ptosis of the eyelids, and low-set ears (fig. 5). Such facies occurred in each of the four patients with a positive family history for pulmonary stenosis. In two of these (cases 9 and 16) the siblings presented normal facies, while in the third (case 11), the mother and a male sibling presented facies similar to that of the patient. The mother of case 2 showed such a facies, although in her a typical dome-shaped valve had been found at operation.

**Electrocardiographic Features**

In 14 of the 16 cases the electrocardiogram showed right axis deviation, while in the remaining two the QRS axis was indeterminate.
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In 13 of the 14 with right axis deviation, the mean manifest QRS axis ranged between +170° and +270°, while in the fourteenth case, the one case with coexistent atrial and ventricular septal defects, the QRS axis was +150°. A comparison of the QRS axes in our 16 cases of pulmonary valvular dysplasia was made with that of 87 children having classical pulmonary valvular stenosis (fig. 6). This showed that in only five patients with classic pulmonary valvular stenosis was the QRS axis greater than +170°, whereas most patients with pulmonary valvular dysplasia exhibited at least this marked degree of right axis deviation.

Among the cases of pulmonary valvular dysplasia, right atrial enlargement, manifested by tall, peaked P waves, was observed in nine patients, while in the remaining seven, the P waves were normal.

Electrocardiographic evidence of right ventricular hypertrophy was present in each case of our series, being of the systolic-overload type in 15. A right ventricular diastolic-overload pattern (rSR' in lead V1) was displayed by case 15, in which an atrial septal defect was also present.

Vectorcardiograms were recorded by the Schmitt SVEC III system in 14 patients. Two patterns of right ventricular hypertrophy resulted. The first seen in each of seven patients was characterized by a QRS vector loop which was directed posteriorly, superiorly, and to the right (fig. 7a). These loops tended to be narrow in each plane and, in the frontal plane, assumed clockwise rotation in three and counterclockwise rotation in four. The second vectorcardiographic pattern was observed in each of the remaining seven cases. It was like that usually seen in classical pulmonary valvular stenosis (fig. 7b). In these, the major QRS vector forces were directed anteriorly, inferiorly, and rightward. The QRS loops

![Figure 6](image)

**Figure 6**

QRS axis in 16 patients with pulmonary valvular dysplasia (△) and in 87 children with typical domeshaped pulmonary valvular stenosis (○).

*Figure 7*

Case 12. Vectorcardiogram recorded at half standardization. Narrow QRS vector loops in each plane. Major portion of QRS electrical forces directed posteriorly, superiorly, and rightward. (b) Case 7. Vectorcardiogram recorded at one-fourth standardization. Major portion of QRS forces directed anteriorly, inferiorly, and rightward.
tended to encompass a larger area than that compared to the group with the first pattern.

**Roentgenographic Features**

The details of the roentgenographic and angiocardiographic findings will be given in another report. In 14 of the 16 cases, thoracic roentgenograms revealed the cardiac size to be normal or at the upper limits of normal. Among these 14, only five showed prominence of the main and left pulmonary arteries, while the pulmonary vascular markings appeared normal in 12 and diminished in the remaining two.

In the remaining two cases, one with co-existent atrial and ventricular septal defects (case 4) and the other with associated atrial septal defect, cardiomegaly and increased pulmonary vascular markings were observed.

**Hemodynamic Features**

Right-sided cardiac catheterization and selective right ventriculography were performed in 14 patients.

Pressures were typical for pulmonary stenosis at the valve level in 13 cases. In the fourteenth case (case 3), the systolic pressure in the pulmonary trunk, although above normal, was below the level of that in the right ventricle. In this case, the levels of pressure in the left and right pulmonary arteries were below those in the pulmonary trunk and were interpreted as evidence of peripheral pulmonary arterial stenosis. At necropsy, such lesions were found to involve the origins as well as branches of the left and right pulmonary arteries (fig. 4).

The right ventricular systolic pressures ranged from 76/0 to 190/0 mm Hg. In three, the values were below 100 mm Hg; in six, between 100 and 150 mm Hg; and in the remaining five, greater than 150 mm Hg. The right ventricular end-diastolic pressure was elevated in 11 cases but in no case did it exceed 15 mm Hg.

In six of the 14 patients, a right-to-left shunt occurring at the atrial level was present and, in five cases, this resulted in mild systemic arterial desaturation (89 to 93%). In the sixth patient, a large right-to-left transatrial shunt resulted in a moderate decrease of systemic arterial saturation (78%). A small left-to-right shunt occurred at the atrial level in two cases.

**Figure 8**

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In one of these (case 15), serial cardiac catheterizations performed on three occasions over a 9-year period revealed no significant change either in the magnitude of the left-to-right shunt or in the level of right ventricular pressure, the latter being above normal in each study. In each of the 14 patients in whom right ventriculography was performed, the nature of the pulmonary valve was different from that of classical dome-shaped pulmonary stenosis. "Doming" of the pulmonary valve either was not observed or, if present, the pattern fell short of the classical picture. Thickened pulmonary cusps, especially identifiable during diastole, were seen (fig. 8). In contrast to classical dome-shaped stenosis, no distinct jet could be observed passing through the valve. Post-stenotic dilatation of the pulmonary trunk was observed in some. Infundibular stenosis was not demonstrated, although subpulmonary muscular stenosis was identified at necropsy in two of the cases (cases 2 and 5).

Results of Operation

Thirteen of the patients were subjected to operations for relief of right ventricular outflow obstruction and to correct any coexistent intracardiac anomalies. Five of the 13 patients (cases 2 to 6) died in the early postoperative period, resulting in operative mortality of 38%.

In four of the five who died after operation, the procedure had been carried out under conditions of cardiopulmonary bypass. In each of these four patients, the pulmonary valve was recognized as having three thickened cusps and a small annulus. In two of these (cases 2 and 3) infundibular resection was carried out, but no procedure was done upon the valve. In the other two, no operative procedure was performed either upon the valve or infundibular area; the associated atrial and ventricular septal defects were closed in one (case 4) and an atrial septal defect was closed in the other (case 6). The fifth patient, who died after operation (case 5), was operated on under conditions of inflow occlusion. The pulmonary valve was described as bicuspid and stenotic with a small central orifice although, later, necropsy was to show that the valve possessed three distinct and dysplastic cusps. Cuspid tissue of the valve was incised. In each case in which operative death occurred, the obstruction had not been adequately relieved and the patient died in a state of low cardiac output.

Operation has been performed on eight of the living patients. One of these (case 16) was operated on a second time to relieve residual stenosis.

In six of the eight living patients who had undergone operation, the pulmonary valve was incised (cases 8, 10, 11, 13, 14, and 16). A right ventricular outflow patch crossing the incised pulmonary annulus was placed in the seventh patient (case 9). At operation in the remaining patient (case 12), thickened, circular constricting tissue at the level of the annulus was observed and tissue was removed. Subsequently, histologic examination of this tissue revealed features observed in dysplastic valves. Because of persistent pulmonary stenosis in case 16, a second operation was performed, at which time an outflow patch was placed across the pulmonary annulus with resultant relief in obstruction.

Postoperative cardiac catheterization has been carried out in each of the eight living patients who had undergone operation (fig. 9). The results of these studies show variation from no change to significant relief of the right ventricular outflow obstruction. In two

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**Figure 9**

Results of serial cardiac catheterization in nine patients with pulmonary dysplasia. Case numbers given in parentheses.
of the patients (cases 8 and 16), following pulmonary valvotomy, there was no change in the level of right ventricular pressure. The two patients (cases 9 and 16) in whom outflow patches were inserted and the one (case 12) in whom tissue was removed from the region of the pulmonary annulus showed successful relief of right ventricular obstruction. In the remaining three patients, each of whom underwent pulmonary valvotomy, the right ventricular pressure was decreased only to about half the preoperative pressure, but in two (cases 11 and 14), the right ventricular systolic pressure remained in excess of 50 mm Hg.

Comment

In classical pulmonary stenosis the flexibility of the valve is precluded by fusion of the valvular tissue around a fixed orifice. In contrast, in pulmonary valvular dysplasia the leaflets are not adherent to one another. Yet obstruction occurs for two reasons. In the first place, the leaflets are highly rigid from intrinsic thickening. The second factor is the tendency for pads of tissue to occur in the sinuses. This tissue acts as a barrier to lateral movement of the leaflets during systole.

The histologic features of the dysplastic pulmonary valve are characteristic. The prominent findings are thickening of the spongiosa and, to a lesser extent, other layers of the cusps by primitive mesodermal-type tissue in conjunction with considerable distortion of the normal architectural landmarks.

The histologic features of the dysplastic pulmonary valve are similar to those described by other authors for lesions involving either the pulmonary valve or other valves. Craig reported on 37 cases of endocardial sclerosis, the majority of which revealed histologic alterations in the mitral and aortic valves. This author also described cases of pulmonary valvular stenosis in which the valves appeared similar to those in our cases. In his experience, the dysplastic pulmonary valve was associated either with right ventricular endocardial sclerosis or dysplasia of one or more of the other valves, the tricuspid valve being involved in some cases.

Fadell and Graziani reported on two infants dying with aortic valvular stenosis in whom the histologic appearance of the aortic valve was similar to that found in the pulmonary valve of our cases. These authors proposed the name, "incomplete differentiation of the valve (myxomatosis)" because of the myxomatous appearance of the valvular tissue. In a subsequent report, identifying similar changes in the aortic valve, Davis and associates used the name, "myxoid dysplasia."

When compared to the classic form of pulmonary valvular stenosis, pulmonary valvular dysplasia was associated with a high operative mortality (38%) when simple incision was done. Procedures to be entertained in treating pulmonary valvular dysplasia include excision of tissue, replacement of valve, and insertion of a gusset across the pulmonary orifice.

Since the operative approach should be different in pulmonary valvular dysplasia compared to classical dome-shaped stenosis clinical and surgical distinction is imperative. Our studies showed several features that are prevalent among patients with pulmonary valvular dysplasia which serve to distinguish this condition from the usual type of pulmonary valvular stenosis. The highlights suggesting dysplasia when pulmonary stenosis is present were given in the section dealing with clinical features. These include (1) a family history of pulmonary stenosis, (2) retardation of both height and weight, (3) abnormal facies, and (4) absence of a pulmonary systolic ejection click in the presence of a systolic ejection type of pulmonary murmur. Additional features suggesting pulmonary valvular dysplasia include excessive degrees of right axis deviation and right ventriculographic findings of a markedly thickened pulmonary valve which presents neither "doming" nor a "jet."

We have observed that during operative exposure the initial impression of the surgeon may be faulty. He may consider the valve to be bicuspid, while in fact the valve possesses three dysplastic leaflets.
Addendum

Since the preparation of this manuscript, we have had the opportunity of confirming with necropsy the clinical diagnosis of pulmonary valvular dysplasia in a patient who was not part of the study reported. This patient, who died at the age of 7 days following pulmonary valvotomy, had manifested the following characteristics clinically: pulmonary systolic murmur without ejection click and typical angiographic features of dyplastic pulmonary valve.

Necropsy showed three thickened pulmonary valvular cusps typical of the condition described. In addition, the tricuspid valve showed short chordae and interadherent leaflets, a picture which suggested an anatomic basis for tricuspid insufficiency.

References

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