Calcific Pulmonic Stenosis

By William C. Roberts, M.D., Dean T. Mason, M.D., Andrew G. Morrow, M.D., and Eugene Braunwald, M.D.

SUMMARY

Seven patients are described in whom calcium deposits occurred in stenotic pulmonic valves. In five the calcific deposits were visible on chest roentgenogram. Survival into adulthood and severe pulmonic stenosis appeared to be the prerequisites for the development of large deposits of calcium in this valve.

Additional Indexing Words:

Valvulotomy

Calcific Pulmonic Stenosis

Calcific stenosis of the aortic valve is a well-recognized and relatively common clinical and pathological entity. In contrast, calcific pulmonic stenosis is rarely encountered either at operation or necropsy. We have observed small deposits of calcium in one or more pulmonic valve cusps in patients who lived into adulthood with left-to-right intracardiac shunts, and in older persons with severe pulmonary hypertension secondary to lung disease. Large deposits of calcium have been seen in this valve, however, only in patients with severe pulmonic stenosis. Seven patients with stenotic calcified pulmonic valves are described in the present report. In five the deposits were visible on chest roentgenograms.

Data on Patients

The clinical data on the seven patients with calcific pulmonic stenosis are summarized in table 1. Their ages at the time of study ranged from 21 to 59 years (average, 37); three were women, four men. All seven subjects had associated defects: tetralogy of Fallot in three, complete transposition of the great arteries and ventricular septal defect in two, valvular aortic stenosis and a foramen ovale type atrial septal defect in one, and a patent foramen ovale in one. Cardiac catheterization performed in five patients disclosed systolic pressures in the right ventricle at the level of the systemic arterial pressure in each, ranging from 102 to 172 mm Hg. In the four patients in whom the pulmonic valve was traversed by the catheter, the pulmonary arterial pressures were normal or low, yielding peak systolic pressure gradients between the right ventricle and pulmonary trunk from 88 to 159 mm Hg. Calcium deposits in the pulmonic valves (figs. 1 to 6) were visible on chest roentgenograms in five subjects; their presence in the pulmonic valves was confirmed anatomically in three of them: at valvulotomy (patients T. M., L. L., and G. L.), and at necropsy (patients L. L., and G. L.). Right ventricular angiograms confirmed the presence of severe valvular pulmonic stenosis in six patients. Four patients had subclavian-pulmonary arterial anastomoses; the pulmonic valvular orifice was found to be totally occluded at open pulmonic valvulotomy 13 years later in patient G. L., and at necropsy 19 years later in patient M. F. It is certain that the pulmonic valve atresia in each patient was acquired since the pulmonary trunk had been entered in each patient by a catheter passed from the right ventricle before the anastomoses had been carried out.

One patient (G. L.) died 12 hours following pulmonic valvulotomy and closure of a
Table 1
Clinical and Pathological Data on Seven Patients with Calcific Pulmonic Stenosis

<table>
<thead>
<tr>
<th>Pt.</th>
<th>Age, sex</th>
<th>Associated lesions</th>
<th>PA pressure s/d (mm Hg)</th>
<th>RV pressure s/d (mm Hg)</th>
<th>PA-RV PSG (mm Hg)</th>
<th>Previous Blalock-Taussig anastomosis</th>
<th>PV calcium seen on chest x-ray</th>
</tr>
</thead>
<tbody>
<tr>
<td>T.M.</td>
<td>59F</td>
<td>VSD</td>
<td>16/9</td>
<td>145/9</td>
<td>129*</td>
<td>0</td>
<td>+</td>
</tr>
<tr>
<td>L.L.</td>
<td>46M</td>
<td>VSD</td>
<td>25/4</td>
<td>130/7</td>
<td>105</td>
<td>+</td>
<td>+</td>
</tr>
<tr>
<td>M.F.</td>
<td>35M</td>
<td>VSD; acquired PV atresia; mesosversion</td>
<td></td>
<td></td>
<td></td>
<td>+</td>
<td>Age 16</td>
</tr>
<tr>
<td>G.L.</td>
<td>33M</td>
<td>Valvular AS; ASD; acquired PV atresia</td>
<td>14/4†</td>
<td>102/6†</td>
<td>88†</td>
<td>+</td>
<td>Age 20</td>
</tr>
<tr>
<td>G.B.</td>
<td>25F</td>
<td>VSD; complete transposition</td>
<td>Not entered</td>
<td>117/5</td>
<td></td>
<td>+</td>
<td>Age 16</td>
</tr>
<tr>
<td>J.G.</td>
<td>21M</td>
<td>VSD; complete transposition</td>
<td></td>
<td></td>
<td></td>
<td>+</td>
<td>Age 10</td>
</tr>
<tr>
<td>A.N.</td>
<td>43F</td>
<td>Patent foramen ovale</td>
<td>13/8</td>
<td>172/12</td>
<td>159</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

Abbreviations: PA = pulmonary artery; RV = right ventricle; PV = pulmonic valve; s/d = systolic/diastolic; PSG = peak systolic gradient; VSD = ventricular septal defect.

*Two years postoperatively this gradient was 22 mm Hg (right ventricle, 55/7 and pulmonary trunk, 33/9 mm Hg).

†Catheterization performed at age 20. At repeat study at age 33 the pulmonic valve could not be crossed by the catheter.

Figure 1

Posteroanterior (A) and lateral (B) chest roentgenograms in patient T.M. The arrows demonstrate calcium deposits in the pulmonic valve.
CALCIFIC PULMONIC STENOSIS

Figure 2
Posteroanterior (A) and lateral (B) chest roentgenograms in patient L. L. The calcium deposits in the pulmonic valve are designated by the arrows.

Figure 3
Oblique chest roentgenogram in patient G.L. Calcium in the pulmonic valve is designated by the arrows. The calcium was not apparent on routine posteroanterior and lateral chest roentgenograms.

Discussion
Although several papers on valvular pulmonic stenosis with or without associated malformations have briefly mentioned calcific deposits in this valve, calcific pulmonic stenosis has been the subject of only two previous reports. Northway and Abrams in 1963 described a 40-year-old man with isolated severe valvular pulmonic stenosis (right ventricular pressure, 193/20 mm Hg; pulmonary trunk not entered by catheter) in whom the calcific deposits in the pulmonic valve were seen on chest roentgenograms and their location in the pulmonic valve confirmed at open valvulotomy. These authors believed that infective endocarditis was a contributor to the heavy valvular calcification, although the patient did not have a history of a cardiac infection. Dinsmore and associates in 1966

Circulation, Volume XXXVII, June 1968
Figure 4
Roentgenogram of the heart (left) and photographs of the pulmonic (P.V.) and aortic (A.V.) valves (right) of patient G.L. In addition to being present in the pulmonic valve, calcium deposits also were present in the aortic valve, although of smaller size, and in the pericardium (black arrows). The pulmonic valve (right) was incised at operation shortly before death.

Figure 5
CALCIFIC PULMONIC STENOSIS

The interior of the heart in patient J.G. This man had complete transposition of the great arteries, ventricular septal defect (D. and V.S.D.), and stenosis of the pulmonic valve (P.V.). Deposits of calcium are present in the severely stenotic pulmonic valve seen from above in the insert, and from below at right. The opened left ventricle (L.V.) from which the pulmonary trunk (P.T.) arises is shown at left. R.V. = right ventricle; and A.M.L. = anterior mitral leaflet.

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traverses the stenotic pulmonic valve as in isolated valvular aortic stenosis. In contrast, a runoff lesion in valvular aortic stenosis is uncommon, since the valve of the foramen ovale is on the left side of the atrial septum and associated ventricular septal defects are infrequent. Whether or not infective endocarditis complicating congenital valvular pulmonic stenosis is an important predisposing cause to the calcification is uncertain. None of the seven patients described herein had a history of a cardiac infection, and only two\textsuperscript{7,9} of the 12\textsuperscript{1-9} previously described patients with calcific pulmonic stenosis had a history of infective endocarditis.

Northway and Abrams\textsuperscript{8} described radiographic findings helpful in distinguishing pulmonic valvular and aortic valvular calcific deposits. The pulmonic valve deposits are more anterior and more cephalad than the aortic valve calcific deposits. The pulmonic valve calcification can be most readily demonstrated in the lateral and right anterior oblique projections.

Whereas calcification of the aortic and mitral valves is common, and calcification of the pulmonic valve rare, deposition of roentgenographically visible calcium in the tricuspid valve has not been reported to our knowledge. We have never observed calcific deposits in the tricuspid valve, even at autopsy, in patients with rheumatic heart disease, but have seen very small focal deposits in the tricuspid valve when it has been traumatized by a jet through a ventricular septal defect or has been the site of a healed vegetation. Sautter and associates\textsuperscript{10} described calcific tricuspid valve stenosis in a 75-year-old woman who also had valvular pulmonic stenosis, ventricular septal defect, and a bicuspid but normally functioning aortic valve.

**Conclusion**

Attention is drawn to roentgenographically visible calcium deposits which were observed in severely stenotic pulmonic valves in five patients. Two additional subjects with severe valvular pulmonic stenosis had small deposits of calcium in this valve at autopsy, but the deposits were not large enough to have been seen on chest roentgenograms. Severe valvular pulmonic stenosis and survival into adulthood appear to be necessary prerequisites for the development of calcification of this valve.

**References**

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Circulation. 1968;37:973-978
doi: 10.1161/01.CIR.37.6.973

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

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