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
Aortic valve calcification

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>0</td>
<td>+</td>
</tr>
<tr>
<td>M.F.</td>
<td>35M</td>
<td>VSD; acquired PV atresia; mesoversion</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.

foramen ovale type atrial septal defect. Death appears to have been the result of associated severe valvular aortic stenosis which had not been corrected at operation. Patient M. F. died of progressive right-sided congestive cardiac failure. Patient J. G. (fig. 5) died of complications of severe polycythemia, and patient A. N. (fig. 6) died at the time of pulmonic valvulotomy.

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
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
Circulation, Volume XXXVII, June 1968
CALCIFIC PULMONIC STENOSIS

Figure 6
Severely stenotic pulmonic valve unopened from above (upper) and after opening (below) in patient A.N. A gradient of 159 mm Hg was present across this valve before valvulotomy. Calcium deposits are present in the thickened cusps.

described roentgenographically visible calcium deposits in five subjects with congenital valvular pulmonic stenosis, four of whom had other cardiovascular anomalies. These patients ranged in age from 35 to 49 years (average, 43), and these authors stated that roentgenographically visible pulmonic valve calcification “seems to be limited to patients with congenital pulmonic stenosis who survive to adulthood without surgical treatment.” In addition, these authors suggested that infective endocarditis was “an important predisposing cause of calcium deposition” in their patients, one of whom had a history of cardiac infection in the distant past. Other reports, which only briefly mentioned pulmonic valve calcification, as well as the seven patients described herein, further emphasize the importance of survival into adulthood in the development of calcification of the pulmonic valve. The youngest patient described with roentgenographically visible pulmonic valve calcification was 22 years old.6 Rawson and Doerner3 in 1953 appear to have been the first to describe roentgenographically visible calcification of the pulmonic valve. Their patient was a 47-year-old man with corrected transposition and dextroversion in addition to congenital valvular pulmonic stenosis.

The reason why heavy deposition of calcium salts rarely occurs in the pulmonic valve is unclear. Calcium deposits in the aortic valve are believed to be the result of trauma to a malformed valve occurring over a long period. Aortic valve calcification is unusual before the age of 20 years and is never present at birth. Thus, if a high ejection pressure is all that is necessary to calcify an aortic valve, which has been made abnormal by either congenital malformation or acquired disease, then calcification of a stenotic pulmonic valve, which is not a rare lesion, should also be more frequent. Certainly in severe pulmonic stenosis the right ventricular systolic pressure may equal or exceed the left ventricular pressure. Few of these patients, however, develop calcified pulmonic valves, probably for two reasons: (1) Few live 40 or 50 years, which is the most common age of patients presenting with isolated calcific aortic stenosis. (2) A runoff, for example, patent foramen ovale or ventricular septal defect (tetralogy of Fallot), is common in patients with valvular pulmonic stenosis, and thus not as much blood

---

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.

Circulation, Volume XXXVII, June 1968
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 \(^7, ^9\) of the 12 \(^1, ^9\) previously described patients with calcific pulmonic stenosis had a history of infective endocarditis.

Northway and Abrams \(^8\) 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 \(^10\) 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**


Calcific Pulmonic Stenosis
WILLIAM C. ROBERTS, DEAN T. MASON, ANDREW G. MORROW and EUGENE BRAUNWALD

_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
Copyright © 1968 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/37/6/973

Permissions: Requests for permissions to reproduce figures, tables, or portions of articles originally published in _Circulation_ can be obtained via RightsLink, a service of the Copyright Clearance Center, not the Editorial Office. Once the online version of the published article for which permission is being requested is located, click Request Permissions in the middle column of the Web page under Services. Further information about this process is available in the Permissions and Rights Question and Answer document.

Reprints: Information about reprints can be found online at:
http://www.lww.com/reprints

Subscriptions: Information about subscribing to _Circulation_ is online at:
http://circ.ahajournals.org//subscriptions/