Supravalvar Stenosis of the Pulmonary Artery

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Narrowing of the pulmonary artery at some site distal of the valves is considered a congenital anomaly which is probably related to maternal rubella (Arvidsson et al., 1961; Emmanouilides, Linde, and Crittenden, 1964; Gyllenswärd et al., 1957; Heiner and Nadas, 1958; Rowe, 1963).

Since the embryological genesis of the condition has not yet been elucidated, anatomical classification at present rests entirely on shape and location of the narrowed segment.

Three main types are distinguished (Hudson, 1965; Nieuven et al., 1961; Smith, 1958; Søndergaard, 1954; Wagenvoort, Heath, and Edwards, 1964):

Type I: Single or multiple stenoses of the pulmonary arteries; either in the main pulmonary trunk, its primary branches, or more peripherally.

Type II: Stenosis of the bifurcation of the pulmonary trunk; “coarctation of the pulmonary artery”.

Type III: Membranous stenosis immediately above the pulmonary valve; “supravalvar stenoctic shelf” (Fig. 1).

The diagnostic problems of supravalvar pulmonary stenosis and its importance in surgery are related to its frequent association with other congenital cardiovascular anomalies.

The following associated lesions have hitherto been described:

1. Pulmonary valvar stenosis (Baum et al., 1964; Coles and Walker, 1956; Emmanouilides et al., 1964; Löhre, Loogen, and Vieten, 1961; Luan et al., 1960; Rowe, 1963; Shumacker and Lurie, 1953; Søndergaard, 1954; Vermillion, Leight, and Davis, 1958; Williams, Lange, and Hecht, 1957).

2. Infundibular pulmonary stenosis (Baum et al., 1964; Löhre et al., 1961; Williams et al., 1957).

3. Fallot’s tetralogy (Baum et al., 1964; Löhre et al., 1961; Luan et al., 1960; Søndergaard, 1954).

4. Ventricular septal defect (Eldridge, Selzer, and Hultgren, 1957; Emmanouilides et al., 1964; Löhre et al., 1961; Luan et al., 1960; Rowe, 1963; Vermillion et al., 1958; Williams et al., 1957).

5. Atrial septal defect (Arvidsson, Karnell, and Möller, 1955; Eldridge et al., 1957; Löhre et al., 1961; Søndergaard, 1954; Williams et al., 1957).


8. Total anomalous pulmonary venous return (Eldridge et al., 1957).

From our own material, to be discussed in this paper, we add:

9. Aortic valvar stenosis (2 cases).
10. Aortic insufficiency (1 case).
11. Coarctation of the aorta (1 case).

Paradoxically, the so-called “associated lesions” assume paramount importance in any discussion of the practical aspects of supravalvar pulmonary stenosis. In the first place, the clinical symptomatology and the results of catheterization and angiography may be distorted beyond recognition by the hemodynamic consequences of the associated lesions. Correct diagnosis may thereby become extremely difficult. Secondly, in the surgical management of the associated lesions the presence of supravalvar pulmonary stenosis introduces a number of hazards which make its pre-operative recog-
Atrial septal valvar stenosis
Aortic atresia of insufficiency
Aortic of Coarctation
Ventricular septal
Membranous stenosis
Stenosis
Stenosis of
Stenosis of
OTHER CARDIOVASCULAR ANOMALIES
of TYPE
then be
mation gained
1.-The
patients might
view
In
these problems,
that the
information gained with present diagnostic methods in
14 patients might be useful.

SUBJECTS AND METHODS
Fourteen patients (8 men, 6 women), whose ages
ranged from 6 to 37 years, underwent complete clinical
examination, including a normal 12-lead electrocardio-
gram, vectorcardiography following the Frank system,
and phonocardiography with an Atlas 6-channel
recorder.

Catheterization studies were performed with the
Telco-equipment, using the micromanometer-catheter
for intracardiac pressure recording and phonography.
In all patients right heart catheterization and retrograde
left heart catheterization were applied.
Pulsed cine-angiocardiology was performed by
means of a Philips 9-inch image amplifier and a 35 mm.
Arriflex camera; the original 35 mm. negative film was
copied onto 16 mm. positive film with the aid of a
reduction-printer. The film frequency was set at 50
frames a second (Chaillet, 1965).

RESULTS
The various types of supravalvar pulmonary
stenosis encountered in our 14 patients are sum-
marized in Table I. In 9 of these patients associ-
ated cardiovascular anomalies were diagnosed (Table
II).

By means of left heart catheterization and angi-
ography from the left ventricle we were able to diag-
ose a number of coexisting conditions (aortic valvar
stenosis, aortic insufficiency, coarctation of the
aorta) which, to our knowledge, have not yet been
described with supravalvar pulmonary stenosis.

Auscultation and Phonocardiography. The aus-
cultatory findings in supravalvar pulmonary stenosis
are not very different from those commonly en-
countered in valvar pulmonary stenosis: that is, a
systolic murmur at the basis of the heart to the right
or left of the sternum, often accompanied by a thrill
(Fig. 2).

There are, however, a number of clinical and
phonocardiographic signs, which make one suspect
supravalvar pulmonary stenosis. The pulmonary
component of the second sound ("second pulmo-
nary sound") which is weak or even inaudible in
other types of pulmonary stenosis is quite loud and
may be audible along the whole left sternal edge and
at the apex (Fig. 3). This sign depends on the
pressure just above the pulmonary valve in relation to
right ventricular pressure. Since, in most types of
distal pulmonary artery stenosis, the pressure in the
pulmonary trunk just beyond the valve is normal or
even raised, a normal or loud pulmonary closing
sound is produced. Sometimes a continuous sys-
tolic-diastolic murmur is encountered, which has
occasionally led to confusion with patent ductus
arteriosus (Arvidsson et al., 1955; Eldridge et al.,
1957; Shafter and Bliss, 1959; Grosse-Brockhoff,
1961). The murmur may be found in any location
on the thorax, depending upon the anatomical type
of the distal pulmonary stenosis.

Fig. 1.—The three main types of supravalvar pulmonary
stenosis (see text).
Supravalvar Stenosis of the Pulmonary Artery

Supravalvar Stenosis of the Pulmonary Artery

TABLE III
SPLITTING OF SECOND SOUND IN 12 PATIENTS WITH SUPRAVALVAR PULMONARY STENOSIS

<table>
<thead>
<tr>
<th>No.</th>
<th>Right ventricular pressures (mm. Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Narrow and inconstant splitting (0–50 msec)</td>
<td>6</td>
</tr>
<tr>
<td>18/1; 20/41; 24/2/5; 29/6/0; 40/1/5; 50/0/1</td>
<td></td>
</tr>
<tr>
<td>Constant splitting due to RBBB (60 msec.)</td>
<td>2</td>
</tr>
<tr>
<td>24/0/0; 28/0/2</td>
<td></td>
</tr>
<tr>
<td>Wide splitting (60–80 msec.)</td>
<td>3</td>
</tr>
<tr>
<td>45/1/3; 52/4; 64/0</td>
<td></td>
</tr>
<tr>
<td>Single 2nd sound</td>
<td>1</td>
</tr>
<tr>
<td>120/0</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>12</td>
</tr>
</tbody>
</table>

Wide splitting (60–80 m.sec. or more) of the second sound is generally present in valvar pulmonary stenosis; the presence or absence of this feature in supravalvar stenosis depends on the haemodynamic conditions (Table III).

Electrocardiography and Vectorcardiography. The electrocardiogram and vectorcardiogram reflect the haemodynamic situation. Therefore, with increasing right ventricular pressures, gradual transition from normal up to right ventricular hypertrophy was found (Table IV).

The vectorcardiogram was used to distinguish more accurately between right bundle-branch block, right ventricular preponderance, and right ventricular hypertrophy (Boutkan, 1965; Greshman, 1965) (Fig. 4). The electrocardiogram is

Fig. 2.—External phonocardiogram of a patient with the membranous type of supravalvar stenosis, complicated by an atrial septal defect (see text). Site: second intercostal space, at right sternal edge. Channels: electrocardiogram and carotid tracing as reference curves. Sound recordings with Maasz-Weber filters at nominal frequencies of 35, 70, 140, and 250 c.p.s.

Fig. 3.—External phonocardiogram of same patient as Fig. 2. Site: at the apex. Channels: electrocardiogram as reference curve, sound recordings without filter, with the "earlike" filter (nominal frequency: 140 c.p.s.) and Maasz-Weber filters at 35, 140, and 250 c.p.s. A = Aortic closure. P = Pulmonary closure, which is well marked (see text).
greatly influenced by associated left ventricular anomalies, and one patient, in whom we also diagnosed valvar aortic stenosis, showed clear-cut left ventricular hypertrophy.

Cardiac Catheterization. The characteristic finding on the withdrawal curves is a rise in the pulmonary artery pressure on passage of the supravalvar stenosis (Fig. 5). If the catheter is withdrawn into the right ventricle, the contour of the pressure curve will change in two steps.

At first sight the recognition of supravalvar stenosis on a catheter/pressure record may not seem too difficult, but it may escape detection even where it has been specifically looked for (Agustsson et al., 1962). Stenosis of the membranous type, where the membrane is often situated only 0.5–1.5 cm. distal of the valve, may easily be missed unless the catheter is withdrawn extremely slowly. In addition, in the presence of an important pulmonary valve stenosis, flow may be so reduced that a concomitant peripheral stenosis does not yield a detectable pressure drop (Williams et al., 1957). The presence of pulmonary valvar stenosis with previously undetected supravalvar stenosis has important surgical implications (Thrower, Abelmann, and Harken, 1960).

From our record (Fig. 5) it will be evident that the intracardiac phonogram shows a loud systolic murmur peripheral from the stenosis. As soon as the catheter has been withdrawn past the stenotic site, the murmur disappears. The intracardiac phonogram, in combination with oximetry and selective angiography, may be most helpful in excluding some sources of error. A pressure-gradient may be simulated by withdrawing the catheter from the “wedge” position (Williams et al., 1957). Pressure changes within the pulmonary artery system may be found when the tip of the catheter passes the pulmonary orifice of a patent ductus arteriosus (Levinson et al., 1951). Small pressure gradients between the primary branches and the main pulmonary artery may be found as a “functional gradient” in conditions with a large pulmonary blood flow due to a left-to-right shunt (Zimmerman, 1966).

The haemodynamic data found in our series of 14 patients may be summarized as follows.

<table>
<thead>
<tr>
<th>Electrocardiogram and vectorcardiogram</th>
<th>No.</th>
<th>Right ventricular pressures (mm. Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>3</td>
<td>18/-1; 24/2/5; 29/-6/0</td>
</tr>
<tr>
<td>Right bundle-branch block</td>
<td>2</td>
<td>24/0/6; 28/0/2</td>
</tr>
<tr>
<td>Right ventricular preponderance</td>
<td>4</td>
<td>40/1/5; 45/1/3; 52/4; 64/0</td>
</tr>
<tr>
<td>Right ventricular hypertrophy</td>
<td>1</td>
<td>120/0</td>
</tr>
<tr>
<td>Total</td>
<td>10</td>
<td></td>
</tr>
</tbody>
</table>
Stenosis of one primary branch was found in 5 patients. In 4 of them, the systolic pressure gradient over the stenosis was quite small (range: from 4 to 12 mm. Hg) and the pressure in the right ventricle was not increased. In one patient, a systolic gradient of 32 mm. Hg was found between distal pulmonary artery and main pulmonary trunk; this patient had a raised right ventricular pressure (Table V). Stenosis of both primary branches and/or the main pulmonary trunk was found in 9 patients. In 7 of them (including one patient with the membranous type of stenosis) the right ventricular pressure was raised.

In 2 patients, with uncomplicated bilateral branch stenosis and extremely small gradients across the stenosis, the right ventricular pressure had remained normal (Table VI). It appears, therefore, that the haemodynamic consequences are

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Stenosed branch</th>
<th>Other cardiovascular anomalies</th>
<th>Pulmonary artery: systolic pressure (mm. Hg)</th>
<th>Pulmonary artery: diastolic pressure (mm. Hg)</th>
<th>Right ventricular pressure (mm. Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Right</td>
<td>Ventricular septal defect</td>
<td>19/8</td>
<td>18/6</td>
<td>24/0/6</td>
</tr>
<tr>
<td>2</td>
<td>Left</td>
<td>Ventricular septal defect</td>
<td>23/9</td>
<td>30/4</td>
<td>52/4</td>
</tr>
<tr>
<td>3</td>
<td>Left</td>
<td>Ventricular septal defect</td>
<td>24/8</td>
<td>24/8</td>
<td>29/6</td>
</tr>
<tr>
<td>4</td>
<td>Right</td>
<td>Coarctation</td>
<td>18/12</td>
<td>11/4</td>
<td>24/2/5</td>
</tr>
<tr>
<td>5</td>
<td>Left</td>
<td>Coarctation</td>
<td>22/12</td>
<td>15/4</td>
<td>20/1</td>
</tr>
</tbody>
</table>

TABLE VI

<table>
<thead>
<tr>
<th>Case No.</th>
<th>Site of stenosis</th>
<th>Other cardiovascular anomalies</th>
<th>Pulmonary artery: systolic pressure (mm. Hg)</th>
<th>Pulmonary artery: diastolic pressure (mm. Hg)</th>
<th>Right ventricular pressure (mm. Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Right and left</td>
<td>Secundum type atrial septal defect</td>
<td>9/4</td>
<td>?</td>
<td>94/7/5/4</td>
</tr>
<tr>
<td>2</td>
<td>Right and left</td>
<td>Secundum type atrial septal defect</td>
<td>14/7</td>
<td>17/7</td>
<td>18/6/1</td>
</tr>
<tr>
<td>3</td>
<td>Right and left</td>
<td>Aortic stenosis</td>
<td>27/12</td>
<td>24/11</td>
<td>28/10/2</td>
</tr>
<tr>
<td>4</td>
<td>Right and left</td>
<td>Aortic insufficiency</td>
<td>56/16</td>
<td>50/12</td>
<td>60/18/3</td>
</tr>
<tr>
<td>5</td>
<td>Right and left</td>
<td>Aortic stenosis</td>
<td>18/12</td>
<td>18/8</td>
<td>50/10/1/4</td>
</tr>
<tr>
<td>6</td>
<td>Right and left</td>
<td>Aortic stenosis</td>
<td>—</td>
<td>—</td>
<td>120/0</td>
</tr>
<tr>
<td>7</td>
<td>Right and left</td>
<td>Valvar pulmonary stenosis</td>
<td>21/4</td>
<td>19/6</td>
<td>33/6/4/0</td>
</tr>
<tr>
<td>8</td>
<td>Membranous</td>
<td>Secundum type atrial septal defect</td>
<td>27/16</td>
<td>27/16</td>
<td>45/12/3</td>
</tr>
<tr>
<td>9</td>
<td>Right and left</td>
<td>Atresia of left branch</td>
<td>30/10</td>
<td>—</td>
<td>40/9</td>
</tr>
</tbody>
</table>

* Catheter could not be passed into the pulmonary trunk.
FIG. 6.—Angiocardiogram showing stenosis of right main branch; frontal view projection. 
M = main pulmonary artery; R = right branch.

FIG. 7.—Angiocardiogram showing stenosis of left main branch; left anterior oblique projection. 
M = main pulmonary artery; L = left branch; RV = right ventricle.

FIG. 8.—Angiocardiogram showing atresia of left branch and multiple peripheral stenoses of the right pulmonary artery, with post-stenotic dilatation; frontal projection. 
M = main pulmonary artery; R = right branch.
determined by the anatomical type of the lesion and the pressure-gradient over the stenotic part.

**Selective Angiocardiography.** If the pressure records obtained on catheterization suggest stenosis of a pulmonary branch or at the bifurcation, then the contrast medium should be injected into the main pulmonary trunk (Fig. 6, 7, 8). If the pressure recordings suggest the membranous type of stenosis or stenosis of the main trunk, then the injection should be made into the right ventricle (Fig. 9).

The diagnosis when the angiographic appearances are typical is fairly easy, but quite often it is impossible to discern the stenotic site. In the branch type of stenosis difficulties related to projection may frustrate adequate demonstration. In membranous stenosis, situated close to the pulmonary valves, the angiographic picture may be indistinguishable from valvar pulmonary stenosis.

**DISCUSSION**

Though the first description of supravalvar pulmonary stenosis dates back as far as 1938 (Oppenheimer, 1938), the clinical importance of this anomaly was not fully realized until the advent of modern cardiac surgery (Søndergaard, 1954).

The surgical implications are important. In the immediate post-operative phase after open intracardiac operation for atrial septal defect, total anomalous pulmonary venous connexion, or atrial septal defect, increased resistance to right ventricular outflow constitutes a serious hazard to life. Unfortunately, as we have shown, the congenital anomalies are frequently associated with supravalvar pulmonary stenosis, and in their surgical management pre-operative recognition and eventually correction of the pulmonary artery stenosis may decide the outcome of the operation (Kirklin and Theye, 1963; Rehder, Kirklin, and Theye, 1962; Theye and Kirklin, 1963).

Narrowed segments of the main pulmonary branches may complicate anastomotic procedures involving a systemic artery and a pulmonary artery (Baum et al., 1964).

Since the pre-operative diagnosis of supravalvar pulmonary stenosis may be impossible in the presence of pulmonary valve stenosis because the flow is too small to yield an appreciable pressure gradient, and since unrecognized and uncorrected pulmonary artery stenosis jeopardizes the outcome of pulmonary valve surgery, operations for pulmonary valve stenosis should as a rule be performed with cardiopulmonary bypass (Thrower et al., 1960).

The supravalvar pulmonary stenoses are important in so far as they may occasionally give rise to a continuous murmur which has to be differentiated from patent ductus arteriosus.

If the murmur is only of the systolic type, then it may appear that supravalvar pulmonary stenosis alone accounts for an otherwise unexplained systolic murmur.

Lastly, the interesting observations of Falkenbach et al. (1959) are relevant. They studied the relationship of unilateral pulmonary artery stenosis to pulmonary hypertension. Their work in dogs suggests that while removal of an entire pulmonary main branch does not raise the pressure in the lesser circulation, a rise in pressure is observed after partial occlusion of a main branch. These authors

![Fig. 9.—Angiocardiogram showing serious deformation of the main trunk, the bifurcation, and both main branches; frontal projection. M = main pulmonary artery; PV = level of pulmonary valve; RV = right ventricle.](image-url)
contemplate the possibility of liberation of a hormonal substance from the ischemic lung as the basis of this phenomenon.

At present, however, unilateral branch stenosis in man will generally not require surgical correction and the membranous type and bifurcation type stenosis will have to be treated according to the hæmodynamic situation.

SUMMARY

The clinical and hæmodynamic data in 14 patients with different types of supravalvar pulmonary stenosis are analysed. In 9 patients the condition was associated with other cardiovascular anomalies. In 4 patients left-heart catheterization revealed the presence of aortic valvar stenosis in 2, coarctation of the aorta in 1, and aortic insufficiency in 1 patient. These associated conditions make the clinical symptomatology very complicated and a correct diagnosis can only be made by combining pressure curves, oximetry, intracardiac phonography, and selective angiography. The practical importance of pre-operative diagnosis of supravalvar pulmonary stenosis in the surgical management of the so-called “associated lesions” is discussed.

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REFERENCES


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