Subvalvar Congenital Mitral Stenosis

A. SÁNCHEZ CASCOS, P. RÁBAGO, M. SOKOLOWSKI, AND J. R. VARELA DE SEIJAS

From the Cardiac Department, Fundación Jiménez Díaz, Madrid, Spain

The recent rapid development of cardiac surgery now demands accurate diagnosis of the rarer types of congenital heart disease; the exact anatomy of congenital valve stenosis, for example, must be determined. The occurrence of valvar, subvalvar, and supravalvar forms of either aortic or pulmonary stenosis is well known.

With respect to congenital mitral stenosis, a rarer cardiac anomaly, we are now beginning to distinguish the valvar type, of which we have previously reported two examples (Varela de Seijas et al., 1960), and the supravalvar form (Rogers et al., 1955; Johnson and Dodd, 1957; Manubens, Krovetz, and Adams 1960); this latter is to be distinguished from cor triatriatum in which an aberrant septum divides the left atrium into two different chambers, whereas in supravalvar stenosis a fibrous stenosing ring is placed immediately above the valve. More recently, Shone et al. (1963) have defined a developmental complex comprising four successive obstructions to left heart flow: supravalvular mitral ring, "parachute" subvalvar anomaly, subaortic stenosis, and coarctation of the aorta; all or only some of these four being present in each of the eight cases they reported. The so-called "parachute" mitral valve was present in half. The term "parachute" is used by these authors by analogy with a parachute whose canopy is represented by the valve leaflets, its strings by the chordae, and its harness by the papillary muscle. The fused leaflets and chordae converge on a single papillary muscle and the subvalvar obstruction is formed by the small orifices between the chordae and papillary muscle.

In rarer cases subvalvar mitral obstruction can be due to the existence of fibrous bands, or abnormal, shortened, and fused chordae tendineae (Daoud et al., 1963). In this connexion Moller et al. (1964), studying cases of congenital endocardial fibroelastosis, either "primary" or "secondary" (i.e. associated with aortic stenosis, aortic coarctation, or aberrant coronary artery), found a typical abnormality of the mitral valve, generally causing mitral incompetence; the leaflets are small and the papillary muscles arise higher than usual on the wall of the left ventricle. This position of the papillary muscles, as well as the shortening of the leaflets, chordae, and even of the papillary muscles, produces the mitral incompetence, but it is clear that this mitral anomaly can sometimes be obstructive, giving a new form of subvalvar mitral obstruction.

We report here three cases of submitral stenosis, all of them associated with other cardiac anomalies. One is an example of the "parachute" type of valve; in the second the valve was involved by congenital endocardial fibroelastosis; the third presented chordal obstruction.

### Case Reports

**Case 1.** This 5-year-old girl was born after a normal full-term pregnancy. The family history was without interest. She had previously been asymptomatic except for frequent respiratory episodes, during which she had mild dyspnea and cyanosis. Recently she also complained of mild dyspnea on effort.

Physical examination revealed a well-nourished girl without cyanosis or clubbing. The chest showed pectus carinatum; the apex beat was felt in the 6th left intercostal space. There was physiological splitting of the second sound, but neither an ejection click nor an opening snap was heard. A grade 5 (out of 6) pansystolic murmur was best heard at the left sternal border and a grade 4 murmur in the mitral area filled diastole. The femoral pulses were present. The electrocardiogram (Fig. 1) showed sinus rhythm at 110 a minute, a clear mitral P wave with a normal P-R interval, QRS of 0·11 sec., with a slow end-phase, negative in I and aVL, positive in II, III, and aVF; precordial leads revealed big isodipsic complexes in V3-5. The electrocardiogram was interpreted as indicating left atrial enlargement with possible biventricular overloading.
Subvalvar Congenital Mitral Stenosis

The chest radiograph (Fig. 2) showed increased lung vascularity, hilar dance, and a globular-shaped heart suggestive of biventricular hypertrophy; the left atrium was found to be dilated in the right anterior oblique position. Blood examination revealed mild polycythaemia (5.3 million).

A clinical diagnosis of ventricular septal defect with mitral obstruction (possibly cor triatriatum) was made.

Right heart catheterization (Table I) revealed severe pulmonary hypertension at near systemic levels; the pulmonary wedge pressure was high (21/13, mean 19 mm. Hg). The blood oxygen saturation was increased in the right ventricle and a left-to-right shunt at ventricular level of 43 per cent of the total pulmonary blood flow was calculated. A persistent left superior vena cava was also found. The final hemodynamic diagnosis was ventricular septal defect with left atrial hypertension.

**TABLE I**

**CASE 1: CATHETERIZATION DATA**

<table>
<thead>
<tr>
<th>Chamber</th>
<th>Pressures (mm. Hg)</th>
<th></th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Systolic</td>
<td>Diastolic</td>
<td>Mean</td>
</tr>
<tr>
<td>Right atrium</td>
<td>3 (8)</td>
<td>–1 (2)</td>
<td>0 (6)</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>74 (80)</td>
<td>2 (6)</td>
<td>55</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>73</td>
<td>37</td>
<td>19</td>
</tr>
<tr>
<td>Pulmonary wedge</td>
<td>21 (22)</td>
<td>13</td>
<td>19</td>
</tr>
<tr>
<td>Left atrium</td>
<td>(22)</td>
<td>(14)</td>
<td>(18)</td>
</tr>
<tr>
<td>Left ventricle</td>
<td>(85)</td>
<td>(12)</td>
<td></td>
</tr>
<tr>
<td>Femoral artery</td>
<td>87 (88)</td>
<td>48</td>
<td>70</td>
</tr>
</tbody>
</table>

Figures in parentheses indicate pressures at operation.

The presence of an important ventricular septal defect being recognized, cine-angiocardiology was performed in an attempt to obtain information about the state of the mitral valve. The injection was made into the pulmonary...
trunk. Passage of the opaque medium across the mitral valve appeared to be slow, but mitral stenosis was not conclusively demonstrated.

It was decided to operate to close the septal defect and to explore the mitral valve. At operation no mitral valve obstruction was found, though a clear mitral diastolic gradient was again demonstrated (Table I). When the ventricle was opened no ventricular septum was found, and banding of the pulmonary artery was then performed. The patient died a few hours after the operation.

Necropsy. The external shape of the heart was triangular, with both great vessels coming out from the left side. Three caval veins drained into the right atrium, the left superior vena cava doing so through the coronary sinus. The right atrium and interatrial septum were normal and so was the tricuspid valve. There was a common ventricle with no evidence of a septum. The great vessels arose from it without transposition or inversion; both aortic and pulmonary valves were normal. The left atrium was externally normal but its wall was thick and the endocardium was white and fibrous. The mitral valve appeared normal when viewed from the atrium, but from the ventricle it was seen to be funnel-shaped with fusion of the leaflets. There was a single bifid papillary muscle and from its apices short partially fused chordae tendineae extended towards the leaflets (Fig. 3), only small orifices allowed blood flow from this subvalvar cul-de-sac to the ventricular cavity: the wider of these orifices was a longitudinal slit placed against the left myocardial wall, while on the internal aspect both apices of the papillary muscle were partially fused by a fibrous membrane. The coronary arteries were normal in course and distribution. No ductus arteriosus was found.

The final diagnosis was persistent left superior vena cava, common ventricle, and subvalvar "parachute"-type mitral stenosis.

Case 2. This 7-year-old boy was born a month prematurely after an otherwise normal pregnancy. There was no significant family history. Congenital heart disease was recognized shortly after birth. He started walking at 2 years and the parents noted then that he was dyspnoic on effort. At 5 years, he developed progressive heart failure that did not respond to the treatment given.

When we saw the patient he was undernourished with severe ascites and oedema. The apex beat was in the 6th left intercostal space and the apical thrust suggested left ventricular hypertrophy. A parasternal right ventricular lift was also felt. On auscultation there was a triple rhythm, due to a third sound, and a pansystolic, grade 4, murmur in the mitral as well as in the tricuspid areas with muffled heart sounds. The remainder of the examination was normal, and the femoral pulses were felt normally.

The electrocardiogram (Fig. 1) showed atrial fibrillation with low voltage complexes (not seen in the Figure which is taken from a later record), and signs of predominant right ventricular hypertrophy, possibly associated with left ventricular hypertrophy; big Q waves were present over the right precordium.

Chest radiographs (Fig. 4) showed an enlarged globular heart with pulmonary congestion.
TABLE II
CASE 2: CATHETERIZATION DATA

<table>
<thead>
<tr>
<th>Chamber</th>
<th>Pressures (mm. Hg)</th>
<th></th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Systolic</td>
<td>Diastolic</td>
</tr>
<tr>
<td>Right atrium</td>
<td>14</td>
<td>8</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>47</td>
<td>6</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>33</td>
<td>15</td>
</tr>
<tr>
<td>Pulmonary wedge</td>
<td>17</td>
<td>12</td>
</tr>
<tr>
<td>Left atrium</td>
<td>17</td>
<td>9</td>
</tr>
<tr>
<td>Left ventricle</td>
<td>85</td>
<td>4</td>
</tr>
</tbody>
</table>

atrium had been reached from the right through a presumed atrial septal defect or patent foramen ovale. These data were incomplete in so far as no blood oxygen saturations had been obtained and atrial pressure curves had not been analysed to get evidence of either mitral incompetence or stenosis. It was decided to repeat the cardiac catheterization when the patient’s cardiac failure had been controlled by treatment. He had a good diuresis and after a month was without ascites or oedema. Cardiac catheterization was about to be performed when the patient suddenly died.

**Necropsy.** Only the heart was examined; its external appearance was normal. The right atrium was enlarged and hypertrophied. There was a large ostium secundum atrial septal defect. The right ventricle and pulmonary artery were normal. The left atrium was also normal.

The mitral valve was thick and deformed, but the area of its orifice was normal. Short chordae tendineae crossed under it fusing the papillary muscles, producing an obvious subvalvar stenosis (Fig. 5). The papillary

---

**FIG. 4.—Chest radiograph of Case 2.**

A diagnosis of endocardial fibro-elastosis with mitral incompetence was made.

The patient had been previously catheterized at another hospital and some data had been obtained. These showed (Table II) moderate pulmonary hypertension and high pressure in both atria with a significant left atrial to left ventricular diastolic gradient. The left

**FIG. 5.—The atrial aspect of the mitral valve in Case 2, showing fused chordae below it.**
FIG. 6.—The opened cavity of the left ventricle in Case 2, showing the white colour of the endocardium and the anomalous high implantation of the papillary muscles.

The papillary muscles were both rudimentary arising from the ventricular wall close to the mitral valve in a very high position (Fig. 6) so that a large part of the ventricular chamber lay between their insertion and the apex.

The endocardium was white and fibrous in all chambers particularly in the left ventricle. Microscopically (Dr. Oliva) the endocardium was found to be thickened in all chambers and a great amount of elastic fibre was located in its superficial layers, while collagenous fibres predominated in the deep ones; the myocardium was not involved (Fig. 7).

The final diagnosis was endocardial fibro-elastosis, atrial septal defect, and congenital deformity of the mitral valve producing both mitral incompetence and subvalvular obstruction.

Case 3. This 11-year-old girl was born after a normal full-term pregnancy, and congenital heart disease was diagnosed shortly after birth. The family history was without interest. Physical development was normal, whereas she was mentally retarded because of severe congenital deafness. She had been asymptomatic except for the presence of mild cyanosis with respiratory episodes.

At 7 years she was catheterized at another hospital and a 100 mm Hg pulmonary valvular gradient was found. A pulmonary valvotomy was then performed, the result being apparently good. When we saw the patient she complained of repeated melena; there had been dyspnea and edema. She showed severe mental impairment and bilateral hypacusia.

The apex beat was normal and a systolic thrill was felt at the pulmonary area. A grade 5/6 systolic murmur was best heard high on the left sternal border; heart
Subvalvar Congenital Mitral Stenosis

Right ventricular catheterization (Table III) revealed moderate right ventricular hypertension with a 22 mm. Hg systolic transpulmonary gradient with normal pulmonary pressures; the pulmonary wedge pressure was normal (9/5, mean 7 mm. Hg). The blood oxygen saturation was increased in the right atrium and a left-to-right shunt at atrial level of 37 per cent of the total pulmonary blood was calculated.

The final hemodynamic diagnosis was atrial septal defect with mild pulmonary gradient probably due to the increased pulmonary flow. Twenty-four hours after cardiac catheterization the patient died following an episode of ventricular fibrillation.

TABLE III
CASE 3: CATHETERIZATION DATA

<table>
<thead>
<tr>
<th>Chamber</th>
<th>Pressures (mm. Hg)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>Systolic</td>
</tr>
<tr>
<td>Right atrium</td>
<td>3</td>
</tr>
<tr>
<td>Right ventricle</td>
<td>42</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>20</td>
</tr>
<tr>
<td>Pulmonary wedge</td>
<td>9</td>
</tr>
<tr>
<td>Brachial artery</td>
<td>123</td>
</tr>
</tbody>
</table>

Necropsy. Only the heart was examined. The external shape was normal; the right atrium was normal except for the presence of an ostium secundum type atrial septal defect. The tricuspid valve was normal and the right ventricle presented only moderate hypertrophy of the parietal band of the crista supraventricularis. On the anterior wall of the right ventricle there was a fibrous circular patch, 1 cm. in diameter. The pulmonary valve

Fig. 8.—Chest radiograph of Case 3.

sounds were normal. Blood pressure was normal and femoral pulses were present. The electrocardiogram (Fig. 1) showed sinus rhythm at 90 a minute, right atrial overloading of diastolic type, and diastolic overloading of the right ventricle with an extreme deviation of the AQRS presenting negative complexes in all leads except aVR and V1–2. The chest radiograph (Fig. 8) presented increased lung vascularity and right ventricular hypertrophy. The patient had anæmia (2·5 million) and positive liver function tests, suggesting cirrhosis.

Fig. 9.—Appearance of the mitral valve in Case 3. The arrow points to the fused chordae under the posterior commissure.
was normal having only two small perforations, probably relics of the previous blind valvotomy. The left atrium and ventricle and aortic valve were normal. The mitral valve was apparently normal when viewed from the atrium, but both commissures presented a central raphe towards the papillary muscles, from which parallel chordae emerged towards the leaflets, resembling a strap (Fig. 9); only small peripheral orifices and a 2 cm. central one allowed the blood flow through this subvalvar mitral obstruction. Coronary arteries were normal. The ductus arteriosus was not found.

The final diagnosis was ostium secundum atrial septal defect with mild infundibular obstruction and subvalvar mitral stenosis.

**DISCUSSION**

These three patients have in common only the presence of submitral stenosis. They are reported in order to emphasize the possibility of this type of congenital mitral obstruction and to discuss its anatomical forms.

The first patient had the typical "parachute" mitral valve, as described by Shone and his colleagues (1963), but it lacked the other elements of the syndrome they described (i.e. supramitral stenosis, subaortic stenosis, and coarctation of the aorta). The association in this case with a single ventricle perhaps explains the development of the "parachute" anomaly in this instance and possibly in others. We think that the lack of formation of the septum could account for the anomaly of the papillary muscles; the other examples of "parachute" valve with a complete septum could perhaps be due to a failure of the formation of the septal contribution to the papillary muscles.

The second patient demonstrates that the typical mitral deformity that occurs in endocardial fibroelastosis can also give rise to subvalvar obstruction of a different anatomical type.

Finally, the third case is an example of another form of submitral stenosis, the obstruction being not critical from the haemodynamic point of view.

**SUMMARY**

Details of three patients who had mitral subvalvar obstruction are reported. In one there was a "parachute" deformity of the valve associated with absence of the ventricular septum; the second had a mitral valve deformed by congenital endocardial fibro-elastosis; the third presented fusion of chordae tendineae.

We wish to express our indebtedness to Dr. Dennis Deuchar, of Guy's Hospital, London, who has kindly revised the text, and to Dr. Oliva, of our Pathology Department, for his microscopical study of the second case.

**REFERENCES**


