

The Developmental Complex of "Parachute Mitral Valve," Supravalvular Ring of Left Atrium, Subaortic Stenosis, and Coarctation of Aorta*

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IN THE course of studies on the pathology of obstructive anomalies of the left side of the heart, it became apparent that there was a tendency for four obstructive, or potentially obstructive conditions to coexist. These anomalies were a "parachute" deformity of the mitral valve, supravalvular ring of the left atrium, subaortic stenosis and aortic coarctation (Fig. 1).

In the 8 cases reported herein, 2 exhibited all four conditions; in the remaining 6 cases, at least two of these anomalies coexisted. This suggested that the four anomalies formed a developmental complex, the former 2 cases representing the complex in its complete form and the latter 6 cases representing *formes frustes*. In individual cases certain of these anomalies, although present, were not sufficiently developed to cause significant obstruction (Table 1).

The primary purpose of this report is to define the individual cardiac anomalies that form this complex, and to indicate what combination was present in each of the 8 cases. Brief reference will be made to the clinical manifestations.

DEFINITION OF THE ANOMALIES OF THE COMPLEX

Parachute Mitral Valve: The deformity called "parachute mitral valve" has the usual

two mitral valvular leaflets and commissures but the chordae, instead of diverging to insert into two papillary muscles, converge to insert into one major papillary muscle (Fig. 2 and 3).

The term "parachute" in describing a deformity of an atrioventricular valve was first used, as far as we are aware, in a paper dealing with corrected transposition; one of us (J. E. E.) was co-author.¹ The analogy was suggested by the shape of the deformed valve. The mitral leaflets resemble the canopy of a parachute, the chordae, its shrouds or strings, and the papillary muscle, the harness. The chordae were often short and thick; this, coupled with their convergent papillary insertion, allowed little mobility of the leaflets. The effect was to create a stenotic mitral valve, since the leaflets were held in close apposition. The only effective communication between the left atrium and the left ventricle was through the interchordal spaces. In the aggregate, these spaces did not allow free egress of blood from the left atrium.

The parachute mitral valve, with its chordal and papillary attachments, resembles that localized anomaly termed a "double orifice of the mitral valve." In the latter condition an opening is present within one of the mitral valve leaflets and from the edges of this opening,

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TABLE I
Summary of Pathologic Findings in 8 Cases Studied

Case	Sex	Age (yrs.)	Supravalvular ring	Sub-aortic stenosis	Parachute mitral	Aortic coarctation	Associated anomalies
1	M	2 3/12	Present & functionally significant	Anomalous m. bundle R.V. L. S.V.C.			
2	M	4	Present & functionally significant	Bicuspid aortic valve			
3	F	13 7/12	Present & functionally significant	Aneurysm of memb. sept. c LV-R.A. comm. Bicuspid A.V.			
4	M	22 6/12	Present & functionally significant	Present & functionally significant	Present & functionally significant	Not present	V.S.D. Bicuspid P.V.
5	F	8/12	Present & functionally significant	Present & functionally significant	Present & functionally significant	Not present	Short mitral chordae Endocard. scl. LV Bicuspid AV
6	M	7/12	Present & functionally significant	Present & functionally significant	Present & functionally significant	Not present	Endocard. scl. in L.V.
7	F	3 4/12	Present & functionally significant	Present & functionally significant	Present & functionally significant	Not present	V.S.D. Accessory pouch of Tricuspid valve
8	F	3/12	Present & functionally significant	Bicuspid A.V.			

Key:  Present & functionally significant Present but not functionally significant Not present.

R.V. = right ventricle; L.S.V.C. = left superior vena cava; L.V.-R.A. Comm. = left ventricular-right atrial communication; A.V. = aortic valve; V.S.D. = ventricular septal defect; P.V. = pulmonic valve; Endocard. scl. = endocardial sclerosis (fibroclastosis); L.V. = left ventricle.

chordae descend to the apex of an underlying papillary muscle.

In one instance (Case 3) two papillary muscles were present, but these were immediately adjacent to each other causing convergence of the chordae at their papillary insertions and yielding the effect of a single papillary muscle (Fig. 4).

Supravalvular Ring of the Left Atrium: This entity is a circumferential ridge of connective tissue that arises at the base of the atrial surfaces of the mitral leaflets and protrudes into the inlet of the mitral valve (Fig. 5.) In the fully developed deformity it acts as a stenosing, perforated diaphragm (Fig. 6A). In some cases it protrudes only slightly and causes no obstruction to the egress of blood from the left atrium (Fig. 6B).

A supravalvular ring was present in each of

the 8 cases studied. It was obstructive in 3 and nonobstructive in the remainder.

Subaortic Stenosis: Two types of subaortic stenosis were observed—the muscular and the membranous. The muscular type is characterized by localized protrusion of hypertrophied ventricular septal tissue into the left ventricular outflow tract (Fig. 7B). This type was observed in 5 of the 8 cases studied.

The membranous type is characterized by circumferential endocardial thickening in the left ventricular outflow region (Fig. 7A). In some cases the two types co-existed. In 2 patients (Cases 3 and 4) the zone of endocardial thickening was sufficiently developed to cause obstruction on its own account.

Coarctation of the Aorta: Four of the cases studied had coarctation of the aorta of the clas-

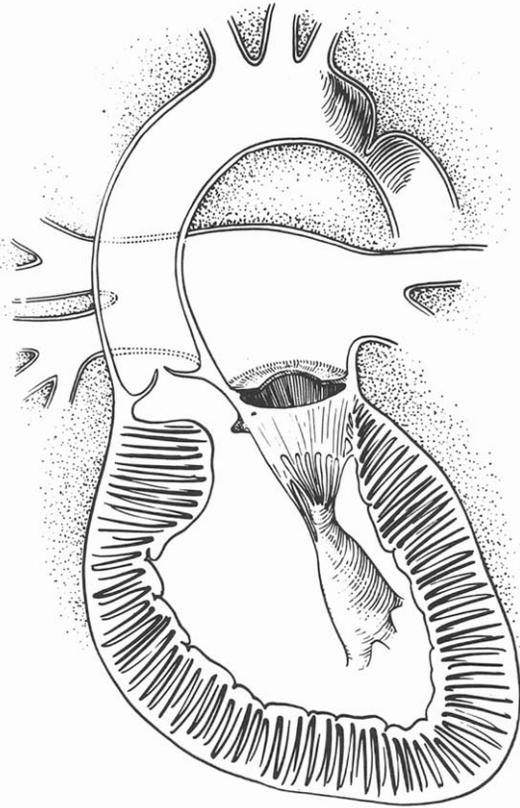


FIG. 1. Diagrammatic representation of the four obstructive anomalies forming the complex: supra-valvular ring of left atrium, parachute mitral valve, subaortic stenosis and coarctation of aorta, in that order, according to the direction of flow of blood.

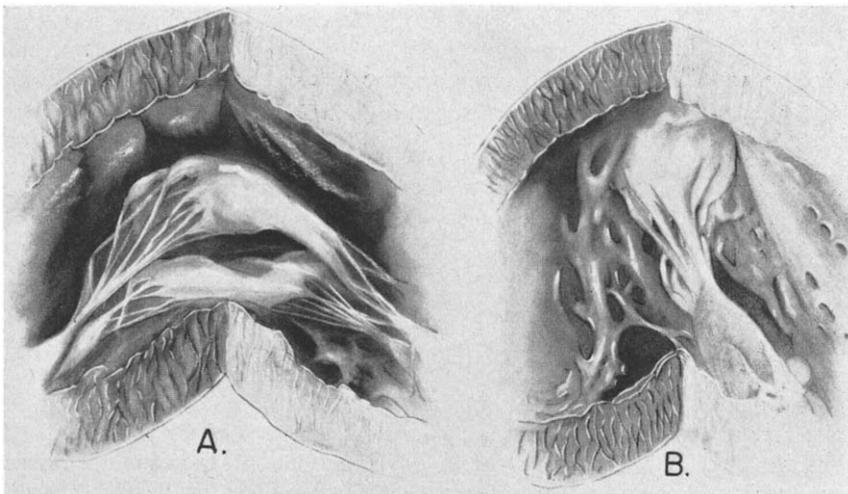


FIG. 2. Artist's representation of mitral valves viewed from below: A, the normal valve. B, the parachute mitral valve. In the normal, the chordae diverge as they insert into two papillary muscles. In the parachute valve, they converge to insert into the single papillary muscle.

sis type,² being located at the junction of the arch and the descending aorta (Fig. 8). In 3 cases the obstruction was significant.

CLINICAL MANIFESTATIONS

Review of the clinical records of the 8 cases showed that the multiplicity of obstructive lesions usually was not suspected clinically and that the clinical manifestations in each case were complex and varied. A brief analysis of the clinical features in relation to the pathologic findings in each case will serve to emphasize the problems involved in clinical diagnosis.

Electrocardiograms were available in 7 of the 8 cases. Although no specific pattern diagnostic of the complex could be identified, tracings from 6* of the cases are presented in Figures 9 and 10 because they may prove useful for comparison in future cases.

CASE REPORTS

CASE 1. Boy, 2 years, 3 months. *Coarctation of the aorta; obstructive supra-valvular ring of left atrium; parachute mitral valve; subaortic stenosis.* (Anomalous muscle bundle of right ventricle was also present.)

Recurrent bouts of congestive cardiac failure had occurred from the age of 5 weeks. "Anginal" attacks accompanied by tachycardia occurred from the age of 9 months. Coarctation of the aorta was resected at the age of 20 months. The child died when he was 2 years and 3 months old at the conclusion of an operation on the mitral valve.

* The electrocardiogram in Case 5 has been published by Rogers and associates.³ (See footnote, Fig. 10.)

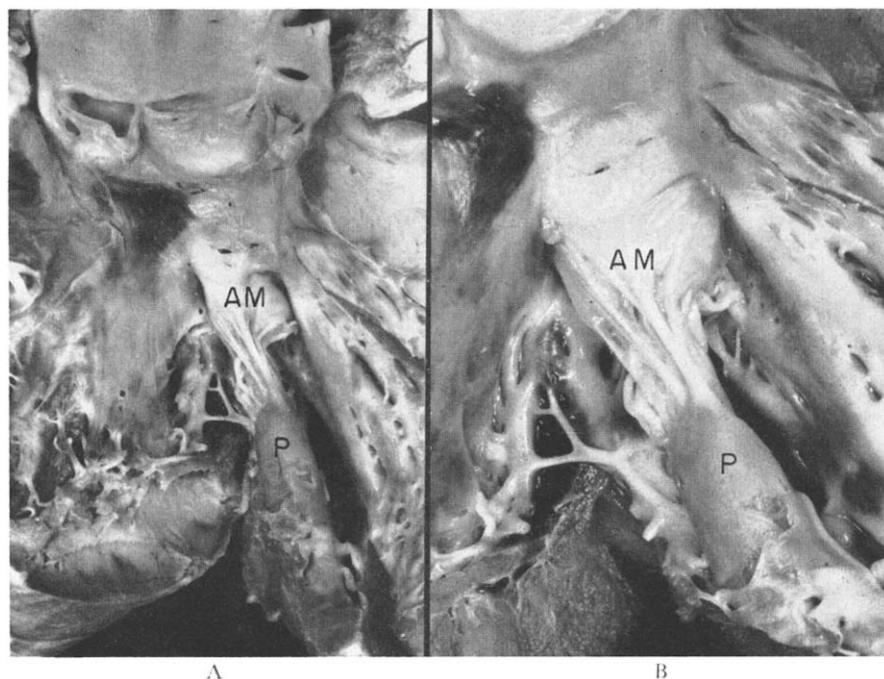


FIG. 3. Case 1. Parachute mitral valve. A, interior of left ventricle and the opened aortic valve. Mitral valve viewed from below. The anterior mitral leaflet (AM) hides most of the posterior leaflet. Chordae of both leaflets insert into the solitary papillary muscle (P). B, close-up view of the mitral valve in the same perspective as shown in A.

The physical findings prior to resection of the coarctation included a grade 3/6 harsh systolic murmur maximal at the third left intercostal space parasternally. There were radiologic findings of enlargement of the left atrium, right ventricle and ascending aorta, together with coarctation of aorta. Coarctation of aorta and endocardial sclerosis were the primary diagnoses. Following resection of the coarctation, the appearance of an apical diastolic rumbling murmur and evidence of pulmonary hypertension provided strong evidence of obstruction at the mitral valve. There was also a harsh, ejection-type systolic murmur at the aortic area.

Angiocardiography showed prolonged opacification of the enlarged left atrium. The electrocardiogram indicated right axis deviation ($\hat{A}QRS + 150^\circ$) and evidence of biventricular hypertrophy with right ventricular systolic (pressure) overload (Fig. 9A). With a preoperative diagnosis of congenital mitral stenosis, a second operation was done.

At operation it was difficult clearly to relate the structures to those of the normal mitral valve. There was no effective means of overcoming the obvious obstruction, and the patient died at the conclusion of the operation. The parachute mitral valve is seen in Figure 3, the supra-ventricular ring of left atrium in Figure 6.

Comment: The aortic coarctation initially dominated the clinical picture. It was only

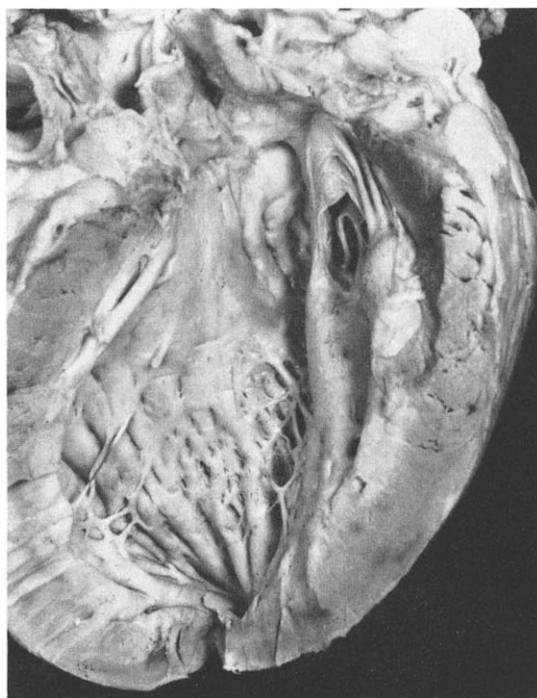


FIG. 4. Case 3. Partial type of parachute mitral valve. Interior of left ventricle. Two papillary muscles are present, but these are immediately adjacent to each other.

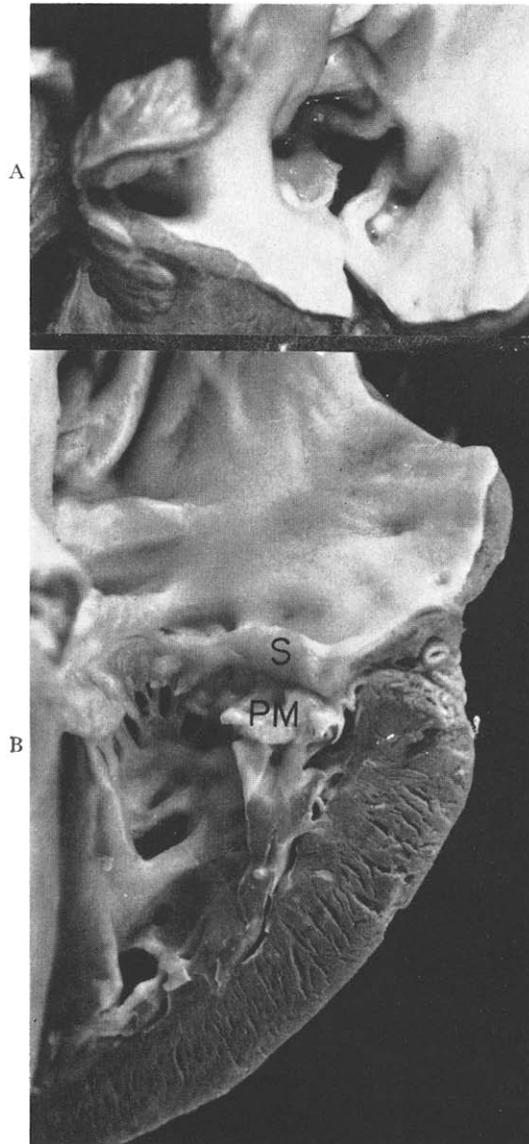


FIG. 5. Case 5. Obstructive supravulvular ring of left atrium. A, the ring is viewed from above. This is genotic and hides the mitral valve, which lies below. The left atrial endocardium is thick, and the wall is hypertrophied. B, longitudinal section through the posterior leaflet of the mitral valve and adjacent structures. Immediately above the posterior leaflet (PM) is the supravulvular ring (S), which attaches to the base of the mitral valve and protrudes into the cavity of the lowermost portion of the left atrium. (From Rogers and associates⁸ with permission of C. V. Mosby Co.)

after resection of the coarctation that obstruction at the mitral valve became clearly apparent. The subaortic stenosis found at necropsy had not been recognized clinically.

CASE 2. Boy, 4 years. *Coarctation of the aorta; parachute mitral valve; subaortic stenosis; nonobstructive*

supravulvular ring of left atrium. (A congenitally bicuspid aortic valve was also present.)

During infancy recurrent attacks of congestive cardiac failure and failure to thrive were noted. Sweating and dyspnea occurred during feedings. A coarctation of aorta was resected at the age of 10 months. The child died at 4 years of age, two days after an operation upon the mitral valve.

The physical findings prior to resection of the coarctation included a grade 2/6 systolic murmur at the second left intercostal space and left sternal border. The second sound was single and accentuated at the pulmonic area. The femoral arterial pulsations were diminished. Blood pressures were 100 mm. Hg systolic in the arms, and 70 mm. Hg in the legs. There were radiologic findings of moderate cardiomegaly with right ventricular enlargement and of severe left atrial enlargement with normal pulmonary vascular markings. The electrocardiogram showed right axis deviation ($\hat{A}QRS + 100^\circ$) and right ventricular hypertrophy of the systolic (pressure) overload type.

After resection of the coarctation the cardiomegaly and left atrial enlargement persisted, and evidence of obstruction at the mitral valve was indicated by progressive right ventricular enlargement, pulmonary arterial hypertension and the appearance of an apical diastolic rumbling murmur (first detected at age 2 years and 4 months). At age 3 years and 6 months a systolic thrill in the suprasternal notch was noted. The electrocardiogram taken immediately before the second operation is shown in Figure 9B.

With the preoperative diagnosis of congenital mitral stenosis, the patient underwent operation. As in Case 1, it was difficult to identify clearly the components of the mitral valve. However, a commissurotomy was performed, which appeared to relieve the obstruction. The patient died two days after operation.

Comment: In this patient too the aortic coarctation dominated the clinical picture initially. Obstruction at the mitral valve became apparent only after resection of the coarctation. No left ventricular angiocardiographic studies were performed, and the subaortic stenosis found at necropsy was not diagnosed clinically.

CASE 3.* Girl, 13 years and 7 months. *Subaortic stenosis; modified parachute mitral valve; mild aortic coarctation; nonobstructive supravulvular ring of left atrium.* (Also present were a perforated aneurysm of membranous septum with left ventricular-right atrial communication and a mildly stenotic, congenitally bicuspid aortic valve.)

A systolic cardiac murmur was detected when the child was 2 years old, but she was asymptomatic apart from fatigue on exertion. Complete atrio-

* Certain aspects of this case have been reported by Drs. M. J. Levy, R. De Wall and C. W. Lillehei.



FIG. 6. Supravulvar ring of left atrium in 2 cases, obstructive in one and nonobstructive in the other. A, Case 1. The ring is viewed from the left atrial aspect. It has been partially incised at operation. The mitral valve is obscured beneath it. B, Case 3. The ring between the left atrium and the mitral valve is evident though nonobstructive. A considerable portion of the mitral valve is readily apparent from this perspective.

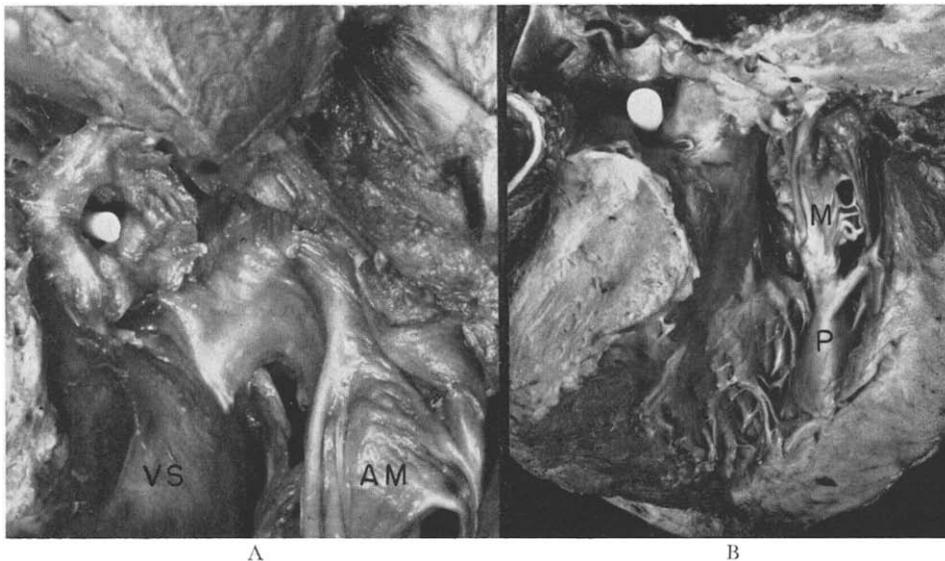


FIG. 7. Case 4. Subaortic stenosis and parachute mitral valve. Ventricular septal defect above the subaortic stenosis is also present. A, the basal aspect of the interior of the left ventricle is exposed. Above a prominent bulge of the ventricular septum (VS) is a fibrous collar which encircles the outflow tract of the left ventricle. The orifice through this area (probe) is severely narrowed. A portion of the anterior leaflet of the mitral valve (AM) is also apparent. B, after the subaortic collar of fibrous tissue illustrated in (A) was excised and the aortic valve opened, the interior of the left ventricle and corresponding valve are exposed. The probe lies in the ventricular septal defect which, in turn, is superior to the zone of subaortic stenosis. The mitral valve (M) shows a typical parachute deformity wherein all the chordae insert into the solitary papillary muscle (P). A stenotic supravulvar ring of the left atrium was also present in this case.

ventricular block occurred following a cardiac operation at the age of 13 years. Death occurred in a Stokes-Adams attack 7 months later.

The physical findings from the age of 4 years included a precordial systolic thrill with a grade 3 to 4/6 harsh, systolic murmur maximal at the third and

fourth left intercostal spaces parasternally and at the aortic area. A short, apical diastolic murmur was heard by some observers. The pulmonic component of the second sound was not accentuated. The femoral arterial pulsations were diminished. Blood pressure in the right arm was 120/85 mm. Hg and

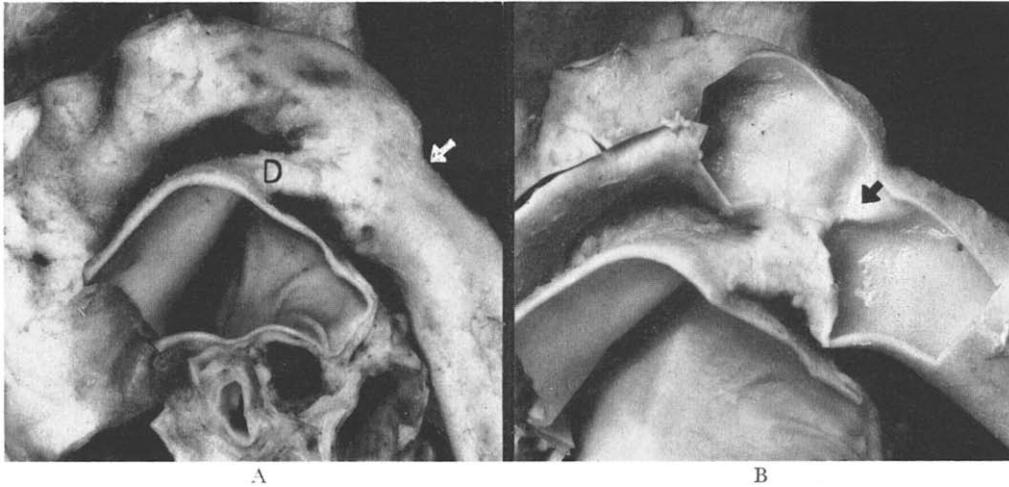


FIG. 8. Case 3. Mild coarctation of aorta. A, external view of the thoracic aorta. Opposite the ligamentum arteriosum (D) is a concave deformity (point of arrow) involving the superior aspect of the aorta. B, interior of the junction of the aortic arch and upper descending aorta. Corresponding to the concavity involving the exterior of the aorta shown in (A) is a curtain-like infolding (point of arrow) which protrudes into the lumen of the aorta. This lesion, characteristic of coarctation of the aorta, caused mild obstruction in this particular instance.

in the right leg, 100/80 mm. Hg. There were *radiologic findings* of marked left atrial enlargement, biventricular enlargement and increased pulmonary vascular markings. The *electrocardiogram* showed a normal QRS axis in the frontal plane and evidence of left ventricular preponderance.

At the age of 6 years the primary diagnoses were ventricular septal defect and mild coarctation of aorta; pulmonary valvular stenosis was considered. At the age of 8 years the patient exhibited a thrill in the suprasternal notch. This was accompanied by a harsh, ejection-type aortic murmur with the paradoxical splitting of the second sound and systolic ejection click of aortic valvular stenosis. The short, apical diastolic murmur was considered that of relative mitral stenosis resulting from the intracardiac left-to-right shunt. At the age of 13 years *selective left ventriculographic studies* revealed a left ventricular-right atrial communication, aortic valvular and subaortic stenosis, and mild aortic coarctation. The electrocardiogram taken prior to operation appears in Figure 9C.

At operation the left ventricular-right atrial communication was closed, and commissurotomy of a stenotic aortic valve and resection of an area of subaortic stenosis were performed. Following operation, the patient exhibited persistent complete atrioventricular dissociation. She died suddenly 7 months following the operation.

Comment: The intracardiac left ventricular-right atrial communication dominated the clinical picture initially. Subsequently definite signs of aortic stenosis developed.

While each of the four anomalies of this com-

plex was represented in this case, one was of major significance. This was the subaortic stenosis which, in conjunction with the left ventricular-right atrial communication, is considered to have been the essential problem to the patient.

In this case, there were two papillary muscles for the mitral valve. Their close apposition contributed to a partial type of parachute deformity (Fig. 4). The nonobstructive ring of left atrium is seen in Figure 6 and the mild aortic coarctation in Figure 8.

CASE 4. Man, 22 years and 6 months. *Parachute mitral valve; obstructive supraavalvular ring of left atrium; subaortic stenosis.* (Also present were a ventricular septal defect and a nonstenotic bicuspid pulmonary valve.)

A systolic murmur was noted at the age of 15 months. Dyspnea and fatigue, associated with mild cyanosis of the lips, occurred on exertion. Following an intracardiac operation, the patient died at the age of 22 years and 6 months.

The *physical findings* at the age of 13 years included left precordial bulging and a systolic thrill accompanied by a grade 4/6, harsh, pansystolic murmur maximal in the third left intercostal space. There was a variable, short, apical diastolic murmur. The second sounds at the aortic and pulmonic areas were equal in intensity. There were *radiologic findings* of left atrial and right ventricular enlargement with increased pulmonary vascular markings. The main pulmonary arterial segment was, however, of normal size. The *electrocardiogram* showed right axis de-

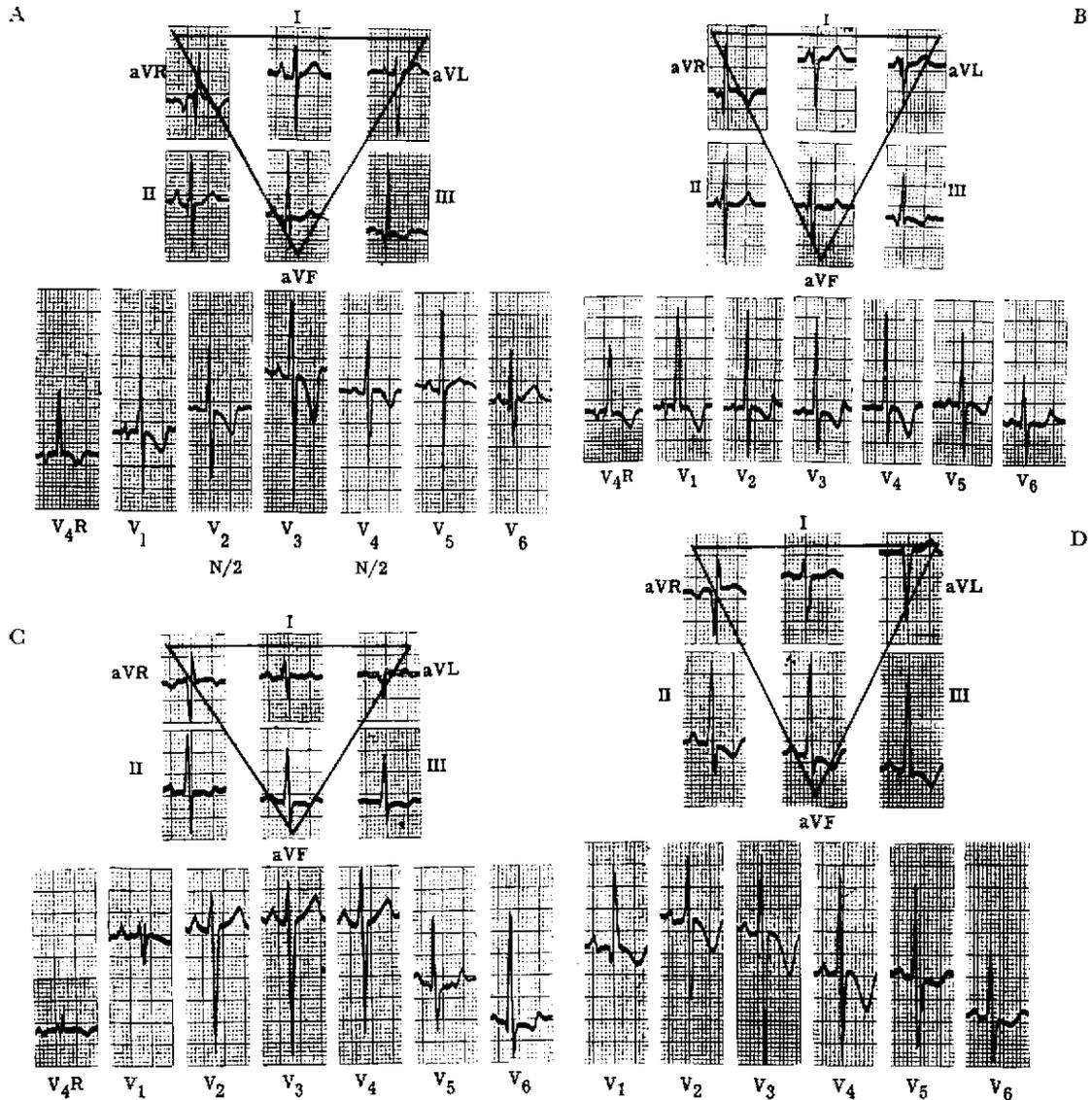


FIG. 9. Electrocardiograms in four cases of the complex. The functionally significant cardiac malformations are indicated. A, Case 1. Supravalvular ring of the left atrium, subaortic stenosis, parachute mitral valve and coarctation of aorta. B, Case 2. Subaortic stenosis, parachute mitral valve and coarctation of aorta. C, Case 3. Subaortic stenosis, left ventricular-right atrial communication. D, Case 4. Parachute mitral valve, supravalvular ring of left atrium, subaortic stenosis and ventricular septal defect.

viation and evidence of right ventricular hypertrophy of the systolic (pressure) overload type.

At this time the diagnoses considered were ventricular septal defect with right-to-left shunt, and a ventricular septal defect with coexisting pulmonic valvular stenosis and patent ductus arteriosus.

Subsequent examination and further studies were refused until the patient was admitted to the hospital at the age of approximately 22 years in a state of congestive cardiac failure associated with atrial flutter. At that time mild cyanosis and digital clubbing were noted. Studies revealed evidence of a large ventricular septal defect with bidirectional

shunt (oxygen saturation of systemic arterial blood 84 per cent), pulmonary arterial hypertension and obstruction at the level of the mitral valve. The electrocardiogram taken prior to operation appears in Figure 9D.

At operation the ventricular septal defect was closed. A mitral commissurotomy was done in an attempt to relieve stenosis of the mitral valve. Irreversible ventricular fibrillation occurred, and after intensive efforts at resuscitation the operation was terminated.

Comment: The clinical picture during childhood was dominated by the ventricular septal

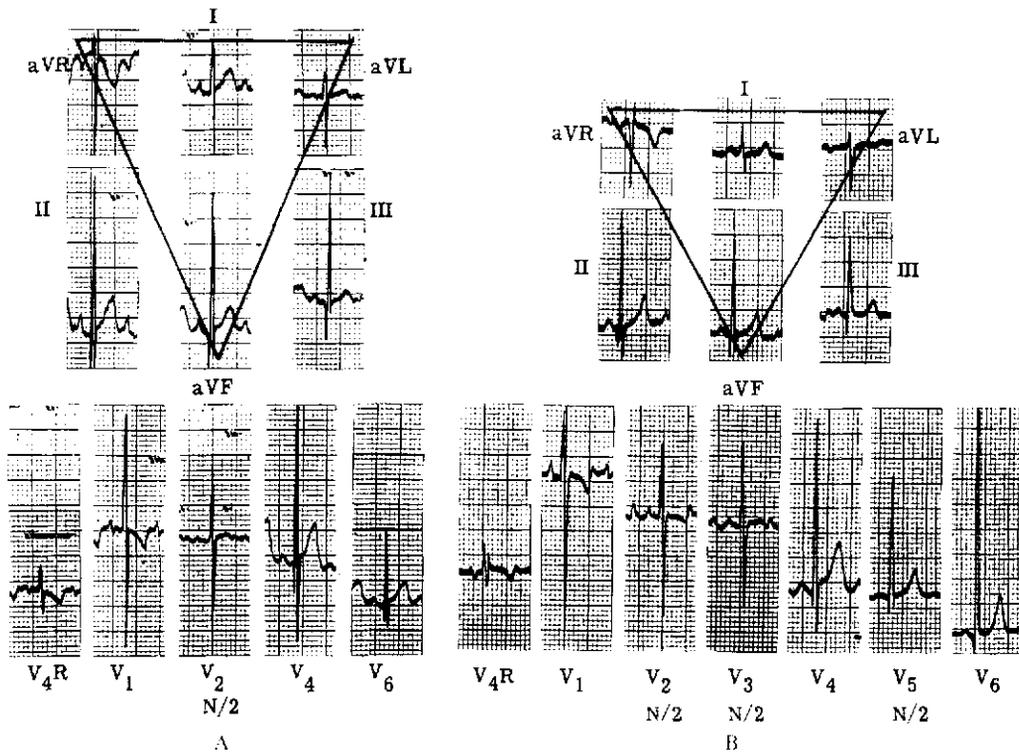


FIG. 10. Electrocardiograms in 2 cases of the complex. Functionally significant cardiac malformations are indicated. A, Case 6. None. B, Case 7. Ventricular septal defect. [Note: The electrocardiogram in Case 5 (supravulvular ring of left atrium and subaortic stenosis) showed a normal QRS frontal plane axis ($+100^\circ$) with evidence of right ventricular hypertrophy of the systolic (pressure) overload type.³ No electrocardiogram was obtained in Case 8.]

defect and the associated pulmonary hypertension. When the patient was an adult, studies revealed obstruction at the mitral valve. The severe subaortic stenosis, located proximal to the large ventricular septal defect, was not suspected during life. Had left ventricular studies been done, it is conceivable that this significant anomaly (Fig. 7) might have been identified. The mitral stenosis recognized clinically was the manifestation of a fully developed parachute mitral valve and an obstructive supravulvular ring of the left atrium (Fig. 7). The commissurotomy had not significantly altered the problem of mitral obstruction, so far as could be ascertained from the necropsy. No aortic coarctation was present.

CASE 5.* Female baby, 8 months. *Obstructive supravulvular ring of left atrium; subaortic stenosis.* (Also present were focal endocardial sclerosis of left ventricle with shortening of mitral chordae tendineae and a congenitally bicuspid aortic valve.)

At the age of 6 months the patient was admitted to the hospital with fever. Rales over the upper right

hemithorax were noted. A grade 3 systolic murmur was heard over the precordium and transmitted to the back. At the age of 7 months she was readmitted because of cyanosis, dyspnea and fever. No cardiac murmur was identified at that time. The second sound was accentuated at the pulmonic area. There were radiologic findings of severe cardiomegaly, a globular-shaped heart and a rounded, upturned cardiac apex. The electrocardiogram showed right axis deviation and evidence of right ventricular hypertrophy of the systolic (pressure) overload type.³ Despite treatment the child died at the age of 8 months in a state of congestive cardiac failure.

Comment: The most prominent features at necropsy were those of an obstructive supravulvular ring of the left atrium (Fig. 5). The other manifestation of the complex which is the subject of this report was muscular subaortic stenosis. While the mitral chordae were abnormally short and thick, there were two normally located papillary muscles.

The thoracic aorta was not available for study in the specimen at our disposal. It is of interest that a bicuspid aortic valve was present. Although this anomaly has a close develop-

* Case previously reported by Rogers and associates.³

mental association with aortic coarctation, it is recognized that a bicuspid aortic valve may occur in the absence of coarctation.

CASE 6. Male baby, 7 months. *Nonobstructive supravalvular ring of left atrium; mild subaortic stenosis.* (Endocardial sclerosis of left ventricle was also present.)

Hospitalized at the age of 5 months for a persistent periumbilical infection and portal venous obstruction, this patient exhibited a grade 2 to 3/6 systolic murmur at the left sternal border. Roentgenograms showed the cardiac size to be at the upper limits of normal, with increased pulmonary vascular markings. There was mild left atrial enlargement. The electrocardiogram showed a normal axis with equivocal evidence of right atrial enlargement and biventricular hypertrophy (Fig. 10A). The patient developed intractable pneumonia and died at the age of 7 months.

Comment: The two components of the complex, a nonstenotic supravalvular ring of the left atrium and a mild membranous subaortic stenosis, probably were of no functional significance in this patient. Their coexistence suggests that this case represents a mild *forme fruste* of the complex.

CASE 7. Girl, 3 years and 4 months. *Mild subaortic stenosis; nonobstructive supravalvular ring of left atrium.* (Also present were a ventricular septal defect and an accessory pouch of tricuspid valve.)

This patient was first seen at 11 months of age because of a problem in feeding and growth. Following percutaneous left ventricular puncture at the age of 3 years and 4 months, cardiac tamponade developed, and the patient died.

The physical findings at the age of 11 months included minimal precordial bulging, a systolic thrill and grade 4/6 harsh systolic murmur maximal in the fourth left intercostal space at the left sternal border. There was a grade 1 to 2 apical rumbling diastolic murmur. The first sound was accentuated at the mitral area. The pulmonic component of the second sound was accentuated. There were *roentgenographic findings* of marked left atrial enlargement, biventricular enlargement and increased pulmonary vascular markings. The *electrocardiogram* showed a normal QRS axis in the frontal plane and evidence of biventricular hypertrophy with left ventricular preponderance. The primary diagnosis was a ventricular septal defect with pulmonary hypertension. Congenital mitral stenosis was considered also.

Studies at the age of 17 months showed evidence of a large left-to-right shunt at ventricular level with severe pulmonary hypertension. Subsequently the physical signs and the electrocardiogram remained essentially unchanged, although increasing cardiomegaly was evident roentgenographically.

At the age of 3 years and 4 months the patient died of cardiac tamponade which followed percutaneous left ventriculography. The electrocardiogram taken prior to these final studies is shown in Figure 10B.

Comment: Though mitral valvular obstruction was considered clinically, no functionally significant disease at this site was demonstrated.

The rudiments of a supravalvular ring of the left atrium which were found, however, are interesting within the framework of this report. Subaortic stenosis of mild muscular type was the only other manifestation of the complex. This case as was Case 6, is a *forme fruste* of the complex.

CASE 8. Female baby, 3½ months. *Coarctation of the aorta; nonobstructive supravalvular ring of left atrium; subaortic stenosis.* (A congenitally bicuspid aortic valve was also present.)

This patient was examined at 1 month of age when no murmur was noted. At 3½ months of age the patient was admitted to the hospital in congestive cardiac failure. A precordial systolic murmur was noted. Roentgenographic examination revealed cardiomegaly. Before an electrocardiogram could be obtained, the patient died.

Comment: Although this patient died before adequate clinical evaluation could be made, examination of the necropsy specimen suggested that coarctation was the most clinically significant portion of the complex. It is conceivable that the subaortic stenosis would not have been noted prior to relief of the coarctation. The supravalvular ring was rudimentary.

DISCUSSION

Among the 8 cases reported as belonging to a developmental complex with four obstructive anomalies of the left side of the heart, a supravalvular ring of the left atrium was present in each. It was obstructive in 3 and nonobstructive in 5. Parachute deformity of the mitral valve was observed in 4 cases. In these, the malformation contributed to (2 cases) or was the sole cause (1 case) of mitral stenosis. In the fourth instance, the deformity was of the partial type and caused no obstruction (Case 3).

Subaortic stenosis present in each of the 8 cases caused significant obstruction to left ventricular outflow in 6 cases, but in only one was this condition suspected clinically. Usually the effects of this malformation were masked by the effects of the coexisting anomaly or anomalies. It is of interest that in the 2 cases in which significant coarctation and mitral obstruction coexisted, the latter condition was not readily ap-

parent clinically until the coarctation was resected.

Re-evaluation of previously reported cases of congenital mitral stenosis would probably reveal some examples of parachute deformity of the mitral valve. Swan and associates⁴ reported "an unusual form of mitral stenosis" in which the mitral valve was described as "unique." "The chordae tendineae were shortened and fused to form a funnel-shaped, firmly fixed, almost solid sheet of tissue that obstructed the orifice of the valve. This anomalous septum was perforated by fifteen, scattered slit-like openings . . . The blended chordae were inserted into the apex of the solitary, double-bellied papillary muscle, which arose from the posterior wall of the left ventricle . . . Viewed from above, the mitral valve resembled the inverted top of a salt shaker." This description, together with Figure 3 of Swan's paper, suggests that this was an example of the parachute mitral valve. This illustration suggests that a supravulvar ring of the left atrium was also present, but this point cannot be established with certainty.

Edwards⁵ reported the case of a 4 day old child with mitral stenosis, ventricular septal defect, tubular hypoplasia of the aortic arch and a bicuspid aortic valve. The features of a parachute mitral valve are shown in his Figure VI-91.

In our basic presentation, we emphasized the association of the parachute deformity of the mitral valve with other obstructive anomalies in the arterial side of the heart and vascular system. In the cases herein reported the basic arrangement of the heart and great vessels was normal. In addition to this series, we have observed one example of a parachute deformity of the mitral valve in a 6 month old child with complete transposition of the great vessels and obstruction of the left ventricular outflow tract. In this case the subpulmonary (left ventricular outflow) obstruction was caused by a membranous ring and anomalous attachment of the anterior mitral leaflet to the ventricular septum. Since two of the four anomalies of the developmental complex described herein were present, it is possible that this case with complete transposition belongs to the complex here reported.

Parachute deformity of an atrioventricular valve may be observed in corrected transposition. Among 14 specimens with corrected transposition Schiebler and associates¹ observed this deformity once in the left atrioventricular

valve and once in the right. El Sayed and associates⁶ illustrated an incomplete form of this anomaly in the right atrioventricular valve of a heart with corrected transposition.

We have had the opportunity of re-examining the specimen in the case reported by Manubens and associates.⁷ Significant obstruction at the mitral valve was caused by a supravulvar ring of the left atrium, but none of the other elements of the complex was present. Coexisting malformations were a ventricular septal defect, extreme dextroposition of the aorta and infundibular stenosis of the right ventricle.

An operation on the mitral valve was performed in 3 of our cases, each having significant obstruction caused both by a supravulvar ring of the left atrium and parachute deformity of the mitral valve. Mitral "commisurotomy" did not relieve the obstruction in any of these 3 cases and does not appear to offer a solution to this complicated problem.

Hypothetically, excision of a supravulvar ring should yield a satisfactory relief of obstruction from this cause. The most feasible method of overcoming the obstruction of a parachute deformity of the mitral valve would appear to be excision of the valve with insertion of a prosthesis.

SUMMARY

A developmental complex is described in which four obstructive anomalies of the left side of the heart and aorta coexist. These anomalies are (1) parachute mitral valve; (2) supravulvar ring of left atrium; (3) subaortic stenosis of either the muscular or membranous type; and (4) coarctation of the aorta.

Eight cases form the basis of this report. In 2 cases, each of the four anomalies was present; all of the other 6 cases represent partial forms of the complex, or *formes frustes*, in that only two or three of the anomalies were present.

The clinical picture is compounded of the effects of the several anomalies of the complex and of the frequent association with still other anomalies, including ventricular septal defect.

REFERENCES

1. SCHIEBLER, G. L., EDWARDS, J. E., BURCHELL, H. B., DUSHANE, J. W., ONGLEY, P. A. and WOOD, E. H. Congenital corrected transposition of the great vessels. A study of 33 cases. *Pediatrics (Suppl.)*, 27: 851, 1961.
2. EDWARDS, J. E., CHRISTENSEN, N. A., CLAGETT, O. T. and McDONALD, J. R. Pathologic considerations in coarctation of the aorta. *Proc. Staff Meet. Mayo Clin.*, 23: 324, 1948.

3. ROGERS, H. M., WALDRON, B. R., MURPHEY, D. F. H. and EDWARDS, J. E. Supravalvular stenosing ring of left atrium in association with endocardial sclerosis (endocardial fibroelastosis) and mitral insufficiency. *Am. Heart J.*, 50: 777, 1955.
4. SWAN, H., TRAPNELL, J. M. and DENST, J. Congenital mitral stenosis and systemic right ventricle with associated pulmonary vascular changes frustrating surgical repair of patent ductus arteriosus and coarctation of the aorta. *Am. Heart J.*, 38: 914, 1949.
5. EDWARDS, J. E. Congenital malformations of the heart. In: GOULD, S. E. *Pathology of the Heart*, ed. 2, p. 388. Springfield, Ill., 1960. Charles C Thomas.
6. EL SAYED, H. et al. Corrected transposition of the great arterial trunks: surgical treatment of the associated defects. *J. Thoracic & Cardiovas. Surg.*, 44: 443, 1962.
7. MANUBENS, R., KROVETZ, L. J. and ADAMS, P., JR. Supravalvular stenosing ring of the left atrium. *Am. Heart J.*, 60: 286, 1960.

