Congenital mitral incompetence and coarctation of aorta

Report of two cases treated surgically¹

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Two patients with congenital mitral incompetence and coarctation of the aorta are presented. One patient had associated patent ductus arteriosus, bicuspid aortic valve, and endocardial fibro-elastosis. The diagnosis in the two patients presented is well established by clinical, laboratory, and surgical findings and also by necropsy examination in one case. It is proposed that the rarity of reported cases in the literature may have resulted from the frequent diagnosis of left ventricular failure in infancy secondary to coarctation, leading to the assumption that a mitral insufficiency murmur, when present, is due to functional regurgitation. Likewise, the murmur may be mistakenly thought to originate from a ventricular septal defect.

The diagnosis of coarctation of the aorta presented no problem in either patient, while detection of the mitral incompetence was difficult. Coarctation of the aorta complicated by pulmonary hypertension in the absence of intracardiac shunt should draw attention to the possibility of associated mitral incompetence. Congestive heart failure, especially after correction of coarctation, was also an indication of possible associated mitral insufficiency.

The two patients were treated by repair of coarctation of the aorta at 3 months and 3 years of age and by mitral valve replacement at the age of 18 months and 5 years, respectively. One patient was in terminal heart failure and died following mitral valve surgery. The other patient benefited from the operation and her case has been followed for over one year. Correction of coarctation of the aorta provided only temporary relief of heart failure. Until both anomalies are corrected response will generally be unsatisfactory.

The aetiology of combined mitral incompetence and coarctation of the aorta can be explained on a congenital basis. Endocardial fibroelastosis of the left ventricle is thought to be secondary to coarctation of the aorta, mitral incompetence, or both.

Congenital mitral incompetence was first described in a 5-year-old girl by Semans and Taussig (1938). This condition, which occurs rarely as an isolated lesion (Berghuis, Kirklin, Edwards, and Titus, 1964), is more frequently associated with anomalies involving the left side of the heart or as part of a syndrome. It has been described as an essential feature of atrioventricular communis, especially with ostium primum defect (Abbott, 1932). It is commonly associated with corrected transposition of the great vessels (Anderson, Lillihei, and Lester, 1957; Helmholz, Daugherty, and Edwards, 1956), ventricular septal defect (Hollman and Hamed,

1965), aberrant origin of the left coronary artery arising from the pulmonary artery (Noren *et al.*, 1964), and endocardial fibroelastosis (Anderson and Kelly, 1956). Congenital mitral incompetence rarely may be associated with congenital aortic stenosis, patent ductus arteriosus (Linde and Adams, 1959), or coarctation of the aorta (Friedberg, 1966).

In reviewing the literature we have found very few descriptions of patients with congenital mitral incompetence and coarctation of the aorta, especially those treated surgically (Flege, Vlad, and Ehrenhaft, 1967; Lillihei, Gott, DeWall, and Varco, 1958; Talner, Stern, and Sloan, 1961). Two patients in whom congenital mitral incompetence was complicated by coarctation of the aorta are

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included in our study. Both patients were treated surgically, initially by repair of the coarctation of the aorta and subsequently by mitral valve replacement.

The purpose of this report is to add these two unusual cases to the literature and to indicate the salient diagnostic features and their surgical implications in the management of this rare combination. Aetiological considerations are also discussed.

CASE REPORTS

CASE 1 A.A., a 3-month-old boy, was admitted on 30 April 1965 because of severe dyspnoea, vomiting, and difficulty in feeding. He was born following a full-term pregnancy and normal labour. Congenital heart disease had not previously been found in this family.

The patient's dyspnoea first appeared at the age of 10 days. Clinical findings of congestive heart failure were recognized and treatment with digoxin resulted in slight improvement. A right heart catheterization had been done at the age of 1 month (Table I). Right ventricular and pulmonary arterial blood pressures were elevated. Blood oxygen saturation studies were consistent with the presence of a ventricular septal defect. Contrast medium injected into the right ventricle opacified a normal right ventricular outflow without evidence of right-to-left shunt. The left atrium was of normal size and emptied rapidly. Contrast medium re-opacified the right ventricle and pulmonary artery from the left ventricle, further evidence of a ventricular septal defect. The opacified aorta appeared narrowed distal to the origin of the left subclavian artery.

The patient had severe respiratory distress with marked substernal retraction. The body weight was 10.9 lb (4.9 kg). The heart rate was 200/minute; the pulses were bounding in the arms but were diminished in both legs. Blood pressure was 130/90 mmHg in the right arm and 80/60 mmHg in the right leg. A

holosystolic murmur (grade 4/6) was heard over the left sternal border and was accompanied by a thrill. Moist râles were heard over both lung bases. Other signs of congestive heart failure were present.

Moderate cardiac enlargement, dilatation of the left atrium, and prominence of the pulmonary arterial markings were seen in radiographs of the chest (Fig. 1A). First-degree A-V block, right axis deviation (AQRS + 110°), and right ventricular hypertrophy were identified on the electrocardiogram.

A diagnosis of coarctation of the aorta was made, and in view of the patient's second episode of heart failure, emergency operation was recommended.

Operation was performed on 30 April 1965. Through a left thoracotomy the aorta was exposed. The coarctation of the aorta was located at the level of a small patent ductus arteriosus. The patent ductus was ligated and divided. The coarctation was resected and end-to-end anastomosis was performed. The lumen of the resected segment of aorta was completely obliterated. Initially the patient improved, but seven days after operation congestive heart failure recurred. He was treated with digitalis and diuretics with some improvement. A residual heart murmur was obvious and increased in intensity during the postoperative period. The murmur was thought to result from a ventricular septal defect, consistent with the heart catheterization findings.

The child was discharged on 26 May 1965 symptomatically improved but taking digitalis.

The patient returned on 3 February 1966, at the age of 1 year. Repeated respiratory tract infections, vomiting, and loss of appetite had been followed by recurrent episodes of loss of consciousness, atonia, and progressive dyspnoea.

The baby was pale and restless. His body weight was 16 lb (7.3 kg). The pulses were readily felt in all extremities. The respiratory rate was 45/minute and there was definite intercostal space retraction. The heart rate was 188/minute. Blood pressure was 110/70 mmHg in the right arm. Jugular venous pressure was elevated and the liver was markedly enlarged.

The apical impulse was in the sixth intercostal space at the anterior axillary line. The right ventricular impulse was hyperdynamic. A holosystolic murmur (grade 3/6) accompanied by a thrill and a diastolic 'flow' murmur (grade 2/6) was heard near the apex.

In a comparative radiological study the heart was as large as before the operation. There was evidence of congestive heart failure and pulmonary oedema. Evidence of right ventricular hypertrophy was still present on the electrocardiogram. The findings during a second heart catheterization are shown in Table I. There was moderate pulmonary arterial hypertension and a normal resting cardiac index. Blood oxygen saturation and dye dilution studies excluded the presence of an intracardiac shunt. Enlargement of all cardiac chambers with marked dilatation of the left atrium was evident in the angiocardiogram.

On the basis of these studies, mitral valve disease, mainly mitral incompetence, was diagnosed. The patient again improved on digoxin and diuretics.

TABLE I
HAEMODYNAMIC DATA: CASE 1

Pressures	Age of Patient		
	1 month		18 months
	Dynamic	Mean	Dynamic Mean
Right atrial	—	3	16/5
Right ventricular	59/0 En.D.	(4)	65/0 En.D.
Pulmonary artery	55/22	40	60/22 En.D.
Pulmonary wedge	—	—	35/25
Left ventricular	—	—	25
Femoral artery	—	—	90/5 En.D.
Pulmonary vascular resistance (Woods units)	—	—	108/75
Cardiac output	—	—	92
Cardiac index	—	—	—
		1.7	1.2
		1.36	3.1
		3.5	—

All pressures in mmHg. En.D. = end-diastolic pressure.

Because of the great risk and the patient's small size, surgery, which seemed indicated, was postponed, and he was discharged on medical treatment.

The third admission was on 24 August 1966, at the age of 18 months. Progressive dyspnoea with congestive heart failure and poor weight gain characterized his course. He was chronically ill and in severe respiratory distress. His body weight was 19.4 lb (8.8 kg). The heart rate was 150/minute and the respiratory rate 50/minute with marked intercostal space retraction; the left chest was obviously deformed. Blood pressure was 110/70 mmHg. There were signs of congestive heart failure. The heart was grossly dilated

to the mid-axillary line and the entire precordium was hyperdynamic. A holosystolic murmur (grade 3/6) accompanied by a thrill and a prominent diastolic inflow murmur was heard near the apex.

Marked cardiomegaly with the left atrium as the predominant chamber was seen on the chest radiograph (Fig. 1B). First-degree A-V block, left axis deviation (AQRS-40°), bi-atrial enlargement, and bi-ventricular hypertrophy were identified on the electrocardiogram.

Data obtained from the third right and left heart catheterization are shown in Table 1. There was marked elevation of the right atrial and right ventricular systolic and end-diastolic blood pressures. The pulmonary artery was not entered. There was no pressure gradient across the aortic valve. The left ventricular end-diastolic pressure was elevated. The cardiac index was normal. Systemic arterial blood was 64% saturated with oxygen; however, breathing 100% oxygen the saturation of the blood increased to 99%. Severe mitral regurgitation was seen by left ventricular angiography. The leaflets of the mitral valve had good mobility. No contrast medium entered the right ventricle.

Severe mitral incompetence was diagnosed and a ventricular septal defect was excluded. Surgical treatment was recommended due to the gravity of the patient's illness and failure of medical treatment to control the congestive heart failure.

Operation was performed on 28 August 1966, using cardiopulmonary bypass at normothermia. Through a median sternotomy the heart was exposed and the

FIG. 1. Comparison of chest radiographs in case 1: (A) before ligation of patent ductus and resection of coarctation; (B) before mitral valve replacement; and (C) on the first day after mitral valve replacement. Note enlargement of the heart at different stages. The prosthetic mitral valve is seen in (C).

FIG. 2. Necropsy specimens from case 1. (A) Demonstrates satisfactory appearance of area of previously resected coarctation: (B) shows extensive endocardial sclerosis of left ventricle. The mitral prosthesis is in position.

left atrium was opened. The mitral valve was grossly insufficient. The posterior leaflet was relatively normal but deficient in substance. The anterior leaflet was cleft, grossly deformed, nodular, and thickened. The chordae tendineae were shortened; a plastic reconstruction was not feasible and the mitral valve was excised and replaced by a No. 1 Kay-Shiley prosthesis. The total perfusion time was 32 minutes.

Microscopic sections of excised valve showed endocardial sclerosis. There were no inflammatory stigmata.

The baby did relatively well in the immediate post-operative period (Fig. 1C). The prosthetic valve sounds were normal and no heart murmur was present. He developed postoperative tachycardia which responded to supplemental digitalis. Approximately 18 hours after surgery the patient had an abrupt cardiac arrest. Resuscitation was attempted without success.

At necropsy the area of the previously treated coarctation was satisfactory (Fig. 2A). There was a bicuspid aortic valve which did not appear to be obstructed or insufficient. The prosthetic mitral valve was in good position and functioning well. The most striking feature was extensive endocardial sclerosis of the left ventricle (Fig. 2B).

Comment The ventricular septal defect, which was suspected following the haemodynamic studies at the age of 1 month and which could not be detected by repeated cardiac catheterization, may be an example of spontaneous closure.

This patient had an unusual combination of five defects: cleft mitral valve in the absence of atrio-ventricular communis, fibroelastosis of the left ventricle, bicuspid aortic valve, coarctation of the aorta, and patent ductus arteriosus (Fig. 3).

CASE 2 R.B., a 3-year-old girl, was first admitted on 5 January 1964 because of marked dyspnoea and fatigue with exertion or crying. Repeated respiratory tract infections had occurred since early childhood. Congenital heart disease was suspected shortly after birth when a heart murmur was detected. The pregnancy was complicated by threatened abortion, and the delivery by asphyxia resulting from twisting of the umbilical cord around the baby's neck. There was no history of congenital heart disease in the family.

The patient did not appear ill. The body weight was 25 lb (11.3 kg). The heart rate was 100/minute. Pulses were bounding in the upper extremities but hardly

FIG. 3. Artist's representation of five pathological features in case 1: cleft mitral valve, bicuspid aortic valve, coarctation of the aorta, patent ductus arteriosus, and fibroelastosis of the left ventricle.

perceptible in the lower extremities. The respiratory rate was 35/minute. Blood pressure was 155/85 mmHg in the left arm and could not be determined in the lower extremities. There were no signs of congestive heart failure. Both left and right ventricular impulses were exaggerated. A holosystolic murmur accompanied by a thrill was heard over the entire precordium and back. A mitral diastolic flow murmur was heard near the apex.

Marked cardiomegaly with enlargement of the left atrium and left ventricle were seen on chest radiographs (Fig. 4A). Pulmonary vascular markings were increased. First-degree A-V block, normal axis (AQRS+75°), left atrial enlargement, and left ventricular hypertrophy were identified on the electrocardiogram.

Right and left heart catheterization findings are shown in Table II. Right ventricular and pulmonary arterial hypertension were present. The left ventricular end-diastolic pressure was elevated. There was a systolic pressure gradient between the ascending aorta and the femoral artery. No evidence of intracardiac shunt was found by blood oxygen saturation or dye dilution study. Severe mitral regurgitation and a dilated left atrium and left ventricle were seen after

TABLE II
HAEMODYNAMIC DATA: CASE 2

Pressure	Pressure (mmHg)	
	Dynamic	Mean
Right atrial	3/0	0
Right ventricular	55/0 En.D.	(6)
Pulmonary artery	55/15	33
Pulmonary wedge	23/12	14
Left atrial	19/13	15
Left ventricular	125/0 En.D.	(14)
Central aortic	155/80	130
Femoral arterial	115/70	80

En.D. = end-diastolic pressure.

injection of contrast material into the left ventricle. A coarctation of the aorta was seen just distal to the origin of the left subclavian artery.

On 12 January 1964 operation was performed through a left thoracotomy. The constricted aortic segment was resected and the aorta repaired with an end-to-end anastomosis. A systolic thrill of mitral incompetence was felt over the left atrium. The patient did well and was discharged 12 days after operation.

The second admission was on 27 November 1966 at the age of 5 years. Improvement after the operation in

1964 had been temporary and was followed by recurrent congestive heart failure which was partially improved by digitalis.

The patient appeared to be chronically ill but not in acute distress. Her body weight was 33 lb (14.9 kg). Peripheral pulses were all present and equal. The blood pressure was 120/80 mmHg and was equal in both upper and lower extremities. The jugular venous pressure was elevated and the liver was enlarged. The apical impulse of the heart was in the sixth intercostal space in the anterior axillary line with exaggerated left ventricular thrust. A mid-diastolic 'flow' murmur was heard over the apex.

Cardiomegaly, marked left atrial and left ventricular enlargement were seen on radiographs of the chest (Fig. 4B). First-degree A-V block, normal axis (AQRS+75°), left atrial enlargement, and left ventricular hypertrophy with strain were demonstrated on the electrocardiogram.

Further surgical treatment was advised as there had been progressive deterioration in spite of medical treatment. Operation was performed on 9 December 1966, using cardiopulmonary bypass at normothermia. Through a median sternotomy the heart was exposed and the left atrium was opened. The mitral valve was grossly insufficient. The annulus was markedly dilated and the leaflets were deficient, thick, and pearly white. The chordae tendineae were thickened and shortened. Plastic reconstruction was not feasible. The valve was excised and replaced by a No. 1 Kay-Shiley prosthesis. The total perfusion time was 29 minutes.

Microscopic sections showed mitral valve sclerosis with interstitial fibrosis of the attached papillary muscles. There was no evidence of inflammation.

The patient did well and was discharged three weeks postoperatively.

She was examined six months after operation. No heart murmurs were heard and the opening and closing sounds of the prosthetic mitral valve were normal. There was marked improvement in exercise tolerance. Improvement in heart size and left ventricular hypertrophy were identified in the radiographs (Fig. 4C) and electrocardiogram.

Comment This patient experienced two prenatal insults which might have produced fetal hypoxic

FIG. 4. Comparison of chest radiographs in case 1: (A) before resection of coarctation; (B) before mitral valve replacement; and (C) 18 days after mitral valve replacement. Note cardiac enlargement at different stages. The prosthetic mitral valve is seen in (C).

stress (Romney, 1966). First, repeated uterine bleeding during pregnancy might have resulted in fetal hypoxia by compromising the placental circulation. Secondly, the umbilical cord was twisted around the neck at birth and resulted in asphyxia at delivery. This might have been associated with a reduced blood flow through the umbilical cord before birth. There is some evidence that coarctation of the aorta may develop, or at least progress, in the neonatal period. It is also possible that severe degrees of coarctation encountered in childhood which may not be compatible with fetal survival have in fact developed after birth (Rudolph, 1966). The relation, if any, between fetal hypoxic stress in the prenatal and postnatal periods and congenital heart disease remains a topic for speculation.

DISCUSSION

A variety of intracardiac malformations occur in combination with coarctation of the aorta. The association of congenital mitral incompetence and coarctation of the aorta has not received much attention in the literature. The remarkably small number of cases reported is indicative of either poor prognosis or rarity of this association. Many cases may be missed by assuming that a mitral insufficiency murmur when present is a functional regurgitation secondary to left ventricular failure produced by the coarctation. In others the murmur may be thought mistakenly to originate from a ventricular septal defect.

The diagnosis of congenital mitral insufficiency and coarctation of the aorta in the two cases reported here is well established by the clinical, laboratory, and surgical findings.

The symptoms that characterized this combination were frequent respiratory infections, dyspnoea on exertion, exercise fatigue, and retarded growth. Heart failure in infancy appeared to be a common complication. Clinical differentiation of mitral incompetence from left-to-right shunts may be difficult and confusion with interventricular septal defect is possible. The systolic murmur in the first case was attributed to a ventricular septal defect, as it was accompanied by a systolic thrill, diastolic flow murmur, and forceful left and right ventricular impulses. A repeat haemodynamic study at 12 months of age proved that no ventricular septal defect was present. Whether ventricular septal defect was anatomically present at the age of 1 month and closed spontaneously, or whether the catheterization data were misinterpreted is unknown.

Characteristically, radiological examination revealed left atrial and left ventricular enlargement with increased pulmonary vascular markings. The electrocardiogram showed first-degree A-V block, left atrial enlargement, and left ventricular hypertrophy. Coarctation of the aorta complicated by pulmonary hypertension, especially in the absence of intracardiac shunt, should draw attention to the possibility of associated mitral incompetence. Mitral incompetence was severe in the two cases presented and was correlated with the severity of the associated pulmonary hypertension.

The diagnosis of coarctation of the aorta presented no problem in either of the two patients in this report. Persistence of congestive heart failure following surgical correction of the coarctation, however, was an indication of the associated mitral insufficiency, which was suspected on clinical grounds. Right and left heart catheterization and angiocardiography confirmed this diagnosis and excluded the presence of other intracardiac defects.

Both patients had a repair of the coarctation of the aorta as a first-stage operation with slight temporary improvement followed by marked deterioration. Before mitral valve replacement was performed at a later stage, the mitral insufficiency produced serious disability and heart failure which did not respond satisfactorily to medical treatment in either patient. Second-stage surgical correction of the mitral insufficiency was performed under unfavourable conditions.

The first patient was severely ill and in spite of medical treatment it was impossible to control his congestive heart failure. Operation was performed as a salvage procedure. The severe fibroelastosis of the left ventricle probably contributed to his deterioration and death. The second patient, in spite of her congestive heart failure, tolerated the operation well and showed marked clinical, radiographic, and electrocardiographic improvement after operation.

Contrary to the preoperative expectation, correction of the coarctation of the aorta and associated patent ductus arteriosus before mitral valve surgery did not result in long-term improvement of cardiac failure. Removal of the aortic obstruction did not appear to reduce the volume of blood regurgitating into the left atrium. Failure to correct the total heart anomaly resulted in an unsatisfactory response.

Although satisfactory results have been reported after plastic reconstruction of congenital mitral insufficiency (Kahn *et al.*, 1967), the severely deformed mitral valves in both of our patients were not amenable to this method of surgical

correction. In spite of the problems which are associated with the use of a prosthetic valve in the pediatric age (Terzaki *et al.*, 1968), mitral valve replacement was the only method which was feasible in both cases.

The aetiology of congenital mitral insufficiency accompanying coarctation of the aorta is not clear. An embryologic association between the two lesions is difficult to explain, but it is possible for the same insult to cause unrelated congenital defects. Shone and his associates described in 1963 the developmental complex of 'parachute mitral valve', supraaortic ring of the left atrium, subaortic stenosis, and coarctation of the aorta. They emphasized the association of the parachute deformity of the mitral valve with other obstructive anomalies in the arterial side of the heart. It is possible that the same factors which predispose to this obstructive mitral valve deformity may result in a deformed insufficient valve.

Another possible explanation is based upon Bremer's postulation (1948) that the expansion of the developing aortic isthmus, located in the relatively stagnant section between the aortic arch and the pulmonary artery, may depend on the volume and rate of blood flow through this section. Diminution of the normal haemodynamic forces operating in the region of the aortic isthmus may play an important role in its failure to expand, resulting in coarctation of the aorta. Congenital mitral incompetence may compromise the forward flow of blood through the aortic arch. Partially deprived of blood current, the isthmus remains small. After the change of circulation accompanying birth, the isthmus normally expands within a short time to full size. Lack of this stimulus to complete normal growth might lead to an arrest of development, predisposing to coarctation of the aorta.

The significance of fibroelastosis in case 1 and possibly in case 2 is more difficult to explain. Fibroelastosis may occur with obstruction to the left ventricular outflow or without associated cardiac malformation. Regardless of the presence or absence of associated cardiac malformation the histological appearance of the endocardium is similar (Moller and Edwards, 1966). Left ventricular overload related to coarctation of the aorta could result in thickening of the left ventricular endocardium and the mitral valve. If the leaflets became thickened and the chordae tendineae shortened, the mitral valve would be insufficient. Surgical exploration, however, revealed that mitral insufficiency in our first patient resulted from a cleft anterior leaflet and a deficient posterior

leaflet. Fibroelastosis which was also present cannot explain the presence of a cleft anterior mitral leaflet in this case. These findings could only result from a developmental defect in the mitral valve. The associated fibroelastosis was probably secondary to coarctation of the aorta, congenital mitral insufficiency, or both.

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ADDENDUM

Case 2 was re-evaluated in October 1972 and the haemodynamics of the circulation are essentially normal.