Congenital Mitral Stenosis

By Charlotte Ferencz, M.D., Arnold L. Johnson, M.D., and F. W. Wiglesworth, M.D.

Nine cases of congenital mitral stenosis are presented, with a review of 34 cases from the literature. A high incidence of associated malformations of the aortic valve, the aorta and the ductus arteriosus is apparent. Clinical findings and results of investigative procedures have been reviewed. Mitral stenosis is responsible for severe circulatory alterations and the development of hypertensive pulmonary vascular changes. This malformation has serious consequences. It is associated with early mortality and renders ineffective the surgical repair of associated coarctation of the aorta or patent ductus arteriosus.

CONGENITAL mitral stenosis has hitherto been considered a rare lesion and only of academic interest, but our attention has recently been directed to the frequent occurrence and practical importance of this malformation. The presence of mitral stenosis in association with other cardiovascular malformations may, on the one hand, alter the hemodynamic sequelae of these associated lesions and, on the other, render their successful surgical correction of no avail. The diagnosis of mitral stenosis in infancy is difficult, and this difficulty is enhanced by the usual association of other abnormalities of the heart or the great vessels.

The present report is based upon a study of nine cases of congenital mitral stenosis encountered in The Children’s Memorial Hospital since 1939 and upon a survey of 34 cases reported in the literature since 1846. In all instances, a significant degree of anatomic stenosis of the mitral valve and a functioning left ventricle were present. Cases of rudimentary mitral valve (“atresia”) and nonfunctioning left ventricle have not been included. It will also be noted that there are no cases of Lutembacher’s syndrome in this series. Mitral stenosis, as an isolated malformation, occurred infrequently and was usually associated with lesions involving the aortic valve and the aorta and only rarely with intracardiac defects (table 1).

In view of the variety of defects associated with the mitral stenosis in our nine cases, it is not feasible to discuss their clinical picture as a group, and it will be necessary to outline briefly the findings in each patient. However, certain observations may be of help in the approach to this difficult diagnostic problem; reference is made to table 2. This group of infants was underweight. Three of them (cases 1, 5, 9) were without symptoms until the age of 9, 8, and 3 months, respectively, when symptoms of acute respiratory distress occurred. Six were in congestive heart failure at some time during the period of observation. The occurrence of pulmonary edema has been given careful consideration. This was an important feature in the clinical course of case 1, and cases 5, 6, 7 and 9 suffered from episodes of respiratory distress which may have represented pulmonary edema. Auscultation was not helpful, for, in only one instance (case 1) was a mitral diastolic murmur heard, and it was transient. In this patient, however, the mitral first and the pulmonic second sounds were abnormally accentuated. The observa-
Table 1.—Incidence of Malformations Associated with Congenital Mitral Stenosis

<table>
<thead>
<tr>
<th>Associated malformation</th>
<th>C.M. Hosp. cases</th>
<th>Literature cases</th>
<th>Total cases</th>
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<tbody>
<tr>
<td>None</td>
<td>—</td>
<td>8</td>
<td>8</td>
</tr>
<tr>
<td>Patent ductus</td>
<td>5</td>
<td>12</td>
<td>17</td>
</tr>
<tr>
<td>Aortic stenosis</td>
<td>4</td>
<td>8</td>
<td>12</td>
</tr>
<tr>
<td>Patent foramen ovale</td>
<td>1</td>
<td>7</td>
<td>8</td>
</tr>
<tr>
<td>Coarctation of aorta</td>
<td>3</td>
<td>4</td>
<td>7</td>
</tr>
<tr>
<td>Aortic valve abnormality</td>
<td>—</td>
<td>5</td>
<td>5</td>
</tr>
<tr>
<td>Hypoplastic aorta</td>
<td>—</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Tricuspid valve abnormality</td>
<td>—</td>
<td>2</td>
<td>2</td>
</tr>
<tr>
<td>Pulmonary valve abnormality</td>
<td>—</td>
<td>1</td>
<td>1</td>
</tr>
<tr>
<td>Defect of ventricular septum</td>
<td>1</td>
<td>1</td>
<td>2</td>
</tr>
<tr>
<td>Double aortic arch</td>
<td>1</td>
<td>—</td>
<td>1</td>
</tr>
<tr>
<td>Fibroelastosis*</td>
<td>8</td>
<td>17</td>
<td>25</td>
</tr>
</tbody>
</table>

* In our own cases we have considered it more likely that the fibroelastosis is a stress effect than a congenital lesion.

The electrocardiogram in the five patients in which it was recorded, showed a pattern of right ventricular hypertrophy, even though, in three of these, the mitral stenosis was associated with lesions which placed a strain on the left ventricle. The evaluation of P-wave changes was confusing, since, in the two cases (1 and 8) in which the P wave was abnormal, it was tall and pointed and interpreted as indicating an increase in the right atrial mass.

The degree of cardiac enlargement observed radiologically was variable. Four patients had extremely large hearts, but, in case 1, a patient in whom there were episodes of pulmonary edema, the heart was only slightly enlarged. Fluoroscopic examination, which was done in three patients (cases 1, 5, 6), revealed no esophageal deviation suggestive of left atrial enlargement.

One patient was studied by cardiac catheterization (case 1). The catheter passed through the ductus arteriosus into the descending aorta and the pressures in the two great vessels were similar. Unfortunately, the pulmonary "capillary" pressure was not recorded.

An angiocardiogram was obtained in case 5 and the presence of mitral stenosis was inferred by the delay in the emptying of the enlarged left atrium.

Case Histories

Case 1. J. H., a 23 month old male child, did well until the age of 9 months when his appetite was observed to fail. At 1 year of age he had the first of three attacks characterized by cough and orthopnea. Between the ages of 9 and 19 months he had not gained any weight and remained very irritable. Our first physical examination at 19 months showed a small, thin child, weighing 15 pounds. The pulses were of normal volume, and the blood pressure was 98/64. The heart was not enlarged. There was no thrill. Auscultation revealed a very loud first sound, an accentuated pulmonic second sound and a moderately loud systolic murmur at the lower left sternal border; a diastolic murmur was thought to be present at the apex, but this could not be determined with certainty. The electrocardiogram showed a pattern of right ventricular hypertrophy. The P waves were tall and peaked in leads II, aVF, and V1. On fluoroscopy, the heart was not definitely enlarged and no selective enlargement of any chamber could be distinguished. There was a diffuse increase in the hilar vascular markings but no evidence of increased blood flow to the lungs.

He continued to do poorly, and, at 26 months of age, was readmitted in severe congestive heart failure which had appeared suddenly and which responded well to therapy. The heart had enlarged considerably since the previous examinations and auscultation revealed an inconstant diastolic rumble at the apex. Periodic episodes of paroxysmal dyspnea occurred, and, following one of these attacks, cyanosis of the left hand and lower extremities was noted. This differential cyanosis was transient and recurrent. On cardiac catheterization, the catheter passed from the pulmonary artery into the descending aorta through a patent ductus. The pressure in the pulmonary artery was of systemic level. At the time of catheterization, there was only a slight amount of venous shunt into the descending aorta, the oxygen saturation of blood in the right radial artery was 92 per cent, and in the descending aorta 85 per cent.

In view of the child’s desperate condition, it was decided to attempt ligation of the patent ductus. There was no evidence of cardiac embarrassment following occlusion of the ductus, and no murmur was present postoperatively. The child appeared to be doing well when, suddenly, on the second postoperative day, he developed marked dyspnea and tachycardia and died.

Anatomic Diagnosis. Mitral stenosis and patent ductus arteriosus (successfully closed at operation).

Comment. The remarkable feature of this case was the marked physical and develop-
mental retardation produced by the cardiac lesions, of which the mitral stenosis was undoubtedly the major one. This occurred in the absence of any significant cardiac enlargement. The occurrence of pulmonary edema and the inconstant diastolic murmur at the apex had suggested the possibility of mitral stenosis, but it was felt that there was not enough evidence to substantiate this diagnosis, and the disappearance of all murmurs following operation lent reassurance to this thought. It must be emphasized that the failure to make the correct diagnosis was in a large measure due to the fact that one is not accustomed to think of mitral stenosis as a major possibility among congenital malformations of the heart.

Case 2. B. R. was an 11 month old male child who had done poorly since birth, there being rapid respiration, difficulty in feeding, weakness and developmental retardation. At 10 months of age, he developed pneumonia and was admitted to another hospital where a diagnosis of patent ductus arteriosus and congestive heart failure was made. He responded well to digitalis and antibiotic therapy and was referred to this hospital for operation. The day before admission, he refused feedings, became feverish and restless and, on arrival, was moribund, showing grayish cyanosis, respiratory distress and evidence of peripheral circulatory collapse. He improved somewhat under energetic therapy. The pulses became forceful and collapsing in type. Blood pressure was 104/30. The heart was enlarged and a machinery-like continuous murmur was audible at the upper left sternal border, accompanied by a thrill. The pulmonic second sound was accentuated. Coarse rales were audible over the entire chest; the liver was just at the costal margin. X-ray films of the chest showed a large heart with increased hilar vascular markings and extensive atelectasis of both lower lobes. Evidence of peripheral circulatory collapse recurred and the baby died within 24 hours of admission.

Anatomic Diagnosis. Mitral stenosis and patent ductus arteriosus.

Comment. On clinical examination, this baby appeared to have the typical findings of a patent ductus with circulatory failure.

Case 3. D. O., a 3 week old male infant, had been observed to have rapid respirations since birth and ate poorly. Since the age of 11 days he had had a cough, and his condition deteriorated rapidly. On admission, he was extremely ill, dyspneic and edematous, but not definitely cyanotic. The lungs were filled with fine moist rales and the liver was enlarged to the level of the umbilicus. The heart was grossly enlarged. There was no thrill. A long, rough systolic murmur of moderate intensity was audible in the pulmonary area and transmitted to the back. An electrocardiogram showed right axis deviation and a QRS pattern of right ventricular hypertrophy within normal limits for the infant's age. The T waves were inverted in leads I, II, aV1, aV6 and in all chest leads. The P waves were not abnormal. X-ray films showed a grossly enlarged heart and increased hilar vascular markings. Despite digitalis and supportive therapy, his condition deteriorated and he died two days after admission. No definite diagnosis of the cardiac lesion had been made.

Anatomic Diagnosis. Congenital mitral stenosis of a mild degree, aortic stenosis, mild coarctation of the aorta, and patent ductus arteriosus (questionably closing).

Case 4. A. C. The history of this 13 month old female child is incomplete, but it appears that her development was poor, and she showed difficulty in breathing. On examination at 11 months of age, there was no cyanosis. The heart was enlarged, and a systolic thrill and murmur were present at the upper left sternal border, the murmur being transmitted to the great vessels of the neck. The liver was slightly enlarged. X-ray films showed marked cardiac enlargement. No definite diagnosis was made. Two months later she was admitted to the hospital, severely ill, and died within one hour.

The late Dr. Maude Abbott examined the specimen and confirmed the anatomic diagnosis of moderate congenital mitral stenosis, aortic stenosis, mild coarctation of the aorta, and patent ductus arteriosus distal to site of coarctation.

Case 5. D. G. was a 25 month old boy who did well until 8 months of age when he had a sudden attack of respiratory distress and, following this, breathed more heavily and faster than before. He also had a chronic cough. During the month preceding admission to the hospital, he became listless and developed swelling of the face. On examination, he was a fairly well developed child in moderately severe congestive failure. The blood pressure in the arms was 120/80. The femoral pulses could not be felt. The heart was enlarged. There was a systolic thrill and a moderately loud, harsh systolic murmur just inside the apex. No diastolic murmur could be elicited. An electrocardiogram showed evidence of marked right ventricular hypertrophy. Fluoroscopy showed a greatly enlarged heart, and the hilar vascular markings were increased. There was no evidence of selective enlargement of the left atrium. The clinical diagnosis of coarctation of the aorta was made and it was believed that this was complicated, probably, by a patent ductus located distal to the site of coarctation, and by pulmonary hypertension, which accounted for the right ventricular hypertrophy observed in the electrocardiogram. An angiocardiogram showed an essentially normal
<table>
<thead>
<tr>
<th>No.</th>
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<th>Age</th>
<th>Weight</th>
<th>Onset sympt.</th>
<th>Cardiac failure</th>
<th>Murmurs:</th>
<th>E.C.G.</th>
<th>Card. enl.</th>
<th>Death</th>
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<td>10 mos.</td>
<td>10 lbs</td>
<td>birth</td>
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<td>-</td>
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<td>+</td>
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<td>Patent ductus, aortic stenosis, coarct. of aorta</td>
<td>13 mos.</td>
<td>?</td>
<td>?</td>
<td>-</td>
<td>-</td>
<td>+</td>
<td>-</td>
<td>Not taken</td>
</tr>
<tr>
<td>6</td>
<td>Aortic stenosis, patent for. ovale</td>
<td>7 weeks</td>
<td>7½ lbs</td>
<td>birth</td>
<td>+</td>
<td>?</td>
<td>+</td>
<td>R.V.H.</td>
<td>Normal</td>
</tr>
<tr>
<td>8</td>
<td>Vent. sept. def., overr. aorta</td>
<td>10½ mos.</td>
<td>14 lbs</td>
<td>birth</td>
<td>-</td>
<td>-</td>
<td>-</td>
<td>R.V.H.</td>
<td>+ Leads 2</td>
</tr>
<tr>
<td>9</td>
<td>Double aortic arch</td>
<td>3½ mos.</td>
<td>9 lbs</td>
<td>3 mos</td>
<td>+</td>
<td>?</td>
<td>-</td>
<td>Not taken</td>
<td>+++</td>
</tr>
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</table>

Abbreviations: R.V.H. = Right ventricular hypertrophy. C.H.F. = Congestive heart failure.
course of the circulation, but dye returning to the left side of the heart was held up in the left atrium and, only in the last film, taken eight seconds after injection, was the aorta visualized. This finding suggested the possibility of an associated mitral stenosis. At operation, the coarctation of the aorta was resected. A small ductus, believed to be closed, entered the aorta just distal to the coarctation. The child died suddenly at the end of operation.

Anatomic Diagnosis. Severe stenosis of the mitral valve. Incomplete subaortic stenosis, coarctation of the aorta (successfully resected), and probe patency of the ductus arteriosus.

Comment. In this case the existence of mitral stenosis with coarctation of the aorta was recognized preoperatively on the basis of the angiocardiographic findings, but the severity of this lesion and its serious consequences were not appreciated.

Case 6. M. L., a 7 week old infant girl, had done poorly since birth, having been great feeding difficulty and failure to thrive. One week prior to admission, she developed a cold and difficulty in breathing. On arrival in hospital she was acutely ill, cyanotic and dyspneic. Râles were heard over the left lung field. The heart was enlarged. A slight thrill was palpable at the upper left sternal border associated with a harsh systolic murmur. The liver was enlarged two fingerbreadths below the costal margin. An electrocardiogram showed a pattern of right ventricular hypertrophy and inverted T waves in all standard leads and in aVL and aVF, and upright T waves in the chest leads. Fluoroscopic examination revealed a grossly enlarged heart and evidence of pulmonary congestion. No area of pulmonary consolidation was identified. The left atrium was not enlarged. The baby responded to oxygen, digitalis and antibiotic therapy and the cyanosis disappeared. Three weeks later, her condition deteriorated somewhat and, on the twenty-sixth hospital day, she suddenly developed marked respiratory distress and died. No definite diagnosis of the cardiac lesion had been made.

Anatomic Diagnosis. Mitral stenosis of slight degree, severe aortic stenosis, patent foramen ovale.

Case 7. H. B., a 16 month old male infant, was brought to the hospital in a moribund state and died within one hour. A detailed history is not available. He was never well, suffering from frequent attacks of "asthma" since early infancy. For several days before admission to the hospital, he had a cold and increasing shortness of breath. As this was apparently a frequent occurrence, the parents showed no concern until he became severely distressed. On examination, he was well developed and well nourished, slightly cyanotic, and in extreme respiratory distress. There was a marked inspiratory stridor and dullness to percussion over the upper lobes of the lungs but no râles. The heart appeared to be enlarged and a soft systolic murmur was heard at the apex. The liver was not enlarged. He was given oxygen, antibiotics and a small dose of adrenaline, but respirations ceased soon after admission and all resuscitative efforts failed. No definite diagnosis was made.

Anatomic Diagnosis. Moderate degree of mitral stenosis and aortic stenosis.

Case 8. C. R., a male infant, was first seen at the age of 10 1/2 months when he was admitted to the hospital and died 12 hours later. He had a "poor color" after birth and, by 1 month of age, was considered to be definitely cyanotic. He always breathed forcefully with difficulty. He fed poorly and his development appeared retarded. For two weeks prior to admission he had a cough, and a diagnosis of bronchopneumonia was made at another hospital. One week before admission to the hospital, he began to have attacks of severe cyanosis in which he became stiff and unresponsive. On the day before admission, he had seven such spells. On examination, he appeared ill and underdeveloped, weighing 14 pounds. He showed moderate cyanosis with early clubbing of the fingers and toes. Respirations were rapid with subcostal indrawing, but there were no râles. The heart sounds were of good intensity and no murmur was present. The liver was 3 cm. below the costal margin. Shortly after admission, he had
### Table 3.—Pathologic Findings

<table>
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<tr>
<th></th>
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<th></th>
<th></th>
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<td>+</td>
<td>+</td>
<td>+++</td>
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<td>+</td>
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<td>--</td>
<td>--</td>
<td>Sl. opaque</td>
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<td>+</td>
<td>+ opaque &amp; thick</td>
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<td>+</td>
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<td>Hypertrophy</td>
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<td>+</td>
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<td>+</td>
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<td>--</td>
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<td>+</td>
<td>+++</td>
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<td>normal</td>
<td>stenosis</td>
<td>stenosis</td>
<td>normal subsaortic shelf</td>
<td>stenosis</td>
<td>stenosis</td>
<td>small but normal</td>
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<tr>
<td>Pulmonary Valve</td>
<td>Sl. large</td>
<td>normal, Sl. sclerosis</td>
<td>normal</td>
<td>normal</td>
<td>large, Sl. sclerosis</td>
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<td>Associated Anomalies</td>
<td>P.D.A.</td>
<td>P.D.A.</td>
<td>Aortic stenosis; mild coarct. of aorta; P.D.A. 7 closing</td>
<td>Aortic stenosis; mild coarct. of aorta; P.D.A. 6 mm. diam. distal</td>
<td>Aortic stenosis; subaortic shelf; coarct. of aorta; P.D.A. probe patent distal</td>
<td>Aortic stenosis; P.F.O. 5 mm. diam.</td>
<td>Aortic stenosis</td>
<td>Basilar I.V.S.D. Sl. overriding of aorta</td>
<td>Double aortic arch with sten. left arch</td>
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<td>Vascular changes</td>
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Grading: + = slight; ++ = moderate; +++ = marked.
an attack of extreme cyanosis with gasping respirations, and distant, slow and irregular heart sounds. The eyes were fixed and the corneal reflex was absent. Oxygen, morphine and adrenaline were given. There was a striking improvement in color shortly after the administration of oxygen was begun with almost complete disappearance of the cyanosis. Respirations remained labored and grunting for some time. A few hours later a similar attack occurred and the infant died in spite of vigorous therapy. The electrocardiogram showed a pattern of marked right ventricular hypertrophy. The P waves in lead II were 3 mm. tall and pointed. In view of the infant's desperate condition, an x-ray film was not made. No definite diagnosis was possible.

Anatomic Diagnosis. Marked mitral stenosis. High basilar ventricular septal defect with slight over-riding of the aorta.

Comment. It is remarkable that this infant had cyanotic attacks similar to those seen in cases of tetralogy of Fallot and he died in such a spell. A dramatic improvement in color followed the administration of oxygen in one of these attacks.

Case 9. C. S. This infant girl was 3\(\frac{1}{2}\) months old. A heart murmur was noted shortly after birth, but the baby did well until the age of 3 months when she developed wheezing respirations and a cough. She became increasingly restless and, on the day of admission, had a severe coughing spell with "froth coming from mouth." On examination, the baby was small, weighing 9 pounds. Respirations were rapid. There were signs of consolidation over the right lung. A murmur was not clearly audible. The liver was enlarged 2 fingerbreaths below the costal margin. X-ray films showed a grossly enlarged heart and evidence of a right upper lobe pneumonia. She received chemotherapy, but her course was downhill and she died on the tenth hospital day. The clinical diagnosis was congenital heart disease with increased blood flow to the lungs, congestive heart failure and pneumonia.

Anatomic Diagnosis. Moderate mitral stenosis and double aortic arch.

Pathologic Findings

The pathologic findings are summarized in table 3. The mitral lesion was characterized by short, thickened, fused and deformed chordae tendineae with endocardial thickening of the apexes of the papillary muscles. In the cases in which there was moderate or marked stenosis, the leaflets were thickened but differed from the leathery scarring of acquired lesions by their semitranslucent, pearly appearance. They were often semicartilagenous in consistency with slightly fluted edges. The chordae were fused into the leaflets. In the two instances of mild stenosis, the leaflets were practically normal, but the chordae were short, fused, and thickened and, on stretching the valves, they appeared somewhat shelf-like and did not flatten out against the ventricular wall. The orifice was usually funnel-shaped, and the circumference of the valve ring was approximately normal except in the instances of marked stenosis.

The endocardium of the left atrium showed well marked thickening in six instances, slight opacification in two, and a normal appearance in one. This change appeared to be correlated with age and with the severity of the mitral stenosis. All of the first six were over 10 months of age and showed moderate to marked stenosis; two were aged 3\(\frac{1}{2}\) and 1\(\frac{1}{2}\) months and showed moderate and slight stenosis, respectively, while the last was 24 days and had slight stenosis. It has been suggested that a stenotic lesion of the mitral or tricuspid valve may lead to "stress" thickening of the atrial endocardium.\(^{32a, 41}\) In this connection, it is of interest that obstructive lesions of the gall bladder give rise to hypertrophy and hyperplasia of the elastic tissue in the wall of this organ.\(^{42}\) The apparent progression of endocardial changes with age in our cases suggests that the duration of the effect of the obstructive lesion played a definite role in the development of the atrial endocardial fibroelastosis. In the light of this evidence, the endocardial changes are unlikely to be of congenital origin. None of the other chambers of the heart showed gross or microscopic evidence of endocardial fibroelastosis, except in one instance (case 7). The fibroelastosis of the left ventricle in this case is believed to be related to the presence of aortic stenosis as there was fairly extensive myocardial atrophy and collapse fibrosis.

There was no strict correlation between the degree of mitral stenosis and dilatation of the left atrium, but there was a reasonable relationship as regards hypertrophy. In comparing the thickness of the right ventricle with that in normal controls, it was evident that right ventricular hypertrophy of varying degree
CONGENITAL MITRAL STENOSIS

Fig. 2. Longitudinal section of the posterior mitral leaflet in case 5. The base of the leaflet is to the right and is normal in appearance. The distal half of the leaflet and the chordae form a confused mass, much of which is loose mesenchymatous tissue. (Masson trichromestain. X 30)

was present in all cases. Left ventricular hypertrophy was present in four cases complicated by aortic stenosis, in one with coarctation of the aorta and a subaortic shelf, and in one case in which the associated lesion was a patent ductus arteriosus.

On microscopic examination, there was no evidence of inflammation or scarring in numerous sections of the heart. In the cases with stenosis of the aortic valve, atrophy of muscle fibers with replacement fibrosis (ischemic) of varying degree was observed. The mitral valves (fig. 2) did not show scarring but presented an abnormal and confused pattern of the layers. Cellular mesenchymatous tissue predominated and was mixed in a most irregular manner with dense collagenous tissue which resembled the ventricularis layer. The basilar part of the leaflet was often normal, the malformation of the layers involving the distal half of the leaflet and the chordae tendineae.

Pulmonary vascular changes were assessed by comparison with normal controls, and no measurements of lumen and wall thickness were made. There were no intimal changes. In the two older cases (1 and 5) medial hypertrophy of the muscular arteries was marked (fig. 3) and was accompanied by mild atherosclerotic changes in the elastic vessels. In case 5, the only apparent cause for these changes was the presence of mitral stenosis. In case 1, although a patent ductus was also present, the severity of the vascular changes was such as to lead one to believe that the presence of marked mitral stenosis played a major rôle in their genesis. In cases 3, 6 and 9 (31/2, 7 and 14 weeks old, respectively) the muscular arteries showed the appearance characteristic of the "fetal state." In the absence of absolute criteria for the end of the fetal state, these changes may be abnormal for these infants. While this cannot be stated with certainty, the presence of right ventricular hypertrophy may be suggestive evidence of this. The muscular arteries showed definite medial hypertrophy in cases 7 and 8 and borderline changes in case 2. In case 4 the pulmonary vasculature appeared entirely normal.

PREVIOUS REPORTS OF CONGENITAL MITRAL STENOSIS IN THE LITERATURE

A search of the literature has yielded 34 cases of congenital mitral stenosis (table 4), present as an isolated malformation in 8 and
Fig. 3. Sections of lung showing muscular artery at about the commencement of a respiratory bronchiole. (H & E stain X 200) (A) Normal control of same age as case 5. (B) Case 5. Note marked medial hypertrophy, small lumen and dense adventitia.

associated with other defects in 26. In the reported cases of Lutembacher's syndrome, a rheumatic origin of the mitral stenosis could not be excluded in any instance and, thus, no such case appears in this series. Of the 34 cases comprising this group, all but one were under 3 years of age and 16 were less than 6 months old.

A review of the histories and physical findings reveals no definite diagnostic criteria but supports certain observations made in connection with our own series. There were eight patients who appeared well in early infancy and had a sudden onset of symptoms between 8 months and 2 1/2 years of age. Cough, dyspnea and congestive heart failure were common. Cyanosis was mentioned in 15 instances, but, at least in some, this may have been terminal or associated with congestive failure. Two patients had differential cyanosis involving the lower extremities, owing to reversed flow through a patent ductus. Accentuation of the heart sounds was rarely clearly described, but two were noted to have had accentuation of the mitral first sound and three of the pulmonary second sound. Five patients were stated to have no murmur. In 12 cases the presence of a diastolic murmur was mentioned; in two of these the murmur was at the left sternal border, in eight the murmur was described as apical or presystolic and in two the location of the murmur was not specified. Electrocardiographic findings were noted in five cases. Three showed a precordial pattern of right ventricular hypertrophy; in two, right axis deviation only was commented upon; P waves were broad and notched in only one instance, tall and pointed in two. Angiocardiographic evidence of mitral stenosis was observed in three cases.

Recent reports emphasize the severity of the pulmonary vascular changes and the disappointing results of otherwise successful surgical correction of coarctation of the aorta and patent ductus because of the presence of mitral stenosis. A mitral valvulotomy was performed in one patient 9 1/2 months of age, reported by Bower and co-workers. These authors have
<table>
<thead>
<tr>
<th>Case No.</th>
<th>Year</th>
<th>Reference</th>
<th>Age</th>
<th>History</th>
<th>Physical findings</th>
<th>Investigation</th>
<th>Pathology</th>
</tr>
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<tbody>
<tr>
<td>1</td>
<td>1846</td>
<td>Smith</td>
<td>21 hrs.</td>
<td>Cyanosis at birth; dyspnea; 'apoplexy'</td>
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<td>P.D.A.—large</td>
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<tr>
<td>2</td>
<td>1885</td>
<td>Ayrolle</td>
<td>10 days</td>
<td>Dyspnea; cyanosis</td>
<td>—</td>
<td>—</td>
<td>?</td>
</tr>
<tr>
<td>3</td>
<td>1894</td>
<td>Carmichael</td>
<td>36 mos.</td>
<td>Dyspnea on feeding; bronchitis; cyanosis</td>
<td>Split sounds; no murmurs</td>
<td>—</td>
<td>Coar. of aorta; P.D.A. into desc. aorta</td>
</tr>
<tr>
<td>4</td>
<td>1900</td>
<td>Cotton</td>
<td>5 days</td>
<td>Cyanosis 1st day</td>
<td>Harsh diast. murmur</td>
<td>—</td>
<td>Defective aortic valve (insufficient); P.D.A. large; P.F.O.</td>
</tr>
<tr>
<td>5</td>
<td>1902</td>
<td>Fisher</td>
<td>15 mos.</td>
<td>Cyanosis at birth; attacks of dyspnea; small size</td>
<td>Syst. murmur; triple sound at apex</td>
<td>—</td>
<td>Coar. of aorta; P.D.A.—pinpoint</td>
</tr>
<tr>
<td>6</td>
<td>1906</td>
<td>Summons</td>
<td>19 mos.</td>
<td>Dyspnea and cyanosis since birth</td>
<td>Thrill; syst. and presyst. murmurs at apex</td>
<td>—</td>
<td>None</td>
</tr>
<tr>
<td>7</td>
<td>1907</td>
<td>Kockel</td>
<td>4 hrs.</td>
<td>—</td>
<td>—</td>
<td>Aort. sten.</td>
<td>—</td>
</tr>
<tr>
<td>8</td>
<td>1911</td>
<td>Fischer</td>
<td>5 wks.</td>
<td>Slight cyanosis; weakness; edema</td>
<td>—</td>
<td>Aort. sten. P.F.O.</td>
<td>—</td>
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<tr>
<td>9</td>
<td>1912</td>
<td>Ludwig</td>
<td>Newborn</td>
<td>Generalized edema</td>
<td>—</td>
<td>P.D.A. — small probe</td>
<td>Thin</td>
</tr>
<tr>
<td>10</td>
<td>1924</td>
<td>Donnelly</td>
<td>3 days</td>
<td>Sudden attack of dyspnea</td>
<td>Syst. murmur at base and back</td>
<td>—</td>
<td>Hypoplastic aorta; P.D.A.—large; P.F.O.</td>
</tr>
<tr>
<td>No.</td>
<td>Year</td>
<td>Name</td>
<td>Age</td>
<td>Symptoms</td>
<td>Signs</td>
<td>Diagnosis</td>
<td>Notes</td>
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<tr>
<td>11</td>
<td>1932</td>
<td>Day</td>
<td>24 mos</td>
<td>Well to 10 mos; dyspnea; cough; poor feeding</td>
<td>Edema; diast. thrill and murmur at apex; short syst. murmur; Accent. P2.</td>
<td>None</td>
<td>Thick in L.A. and R.V.</td>
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<tr>
<td>12</td>
<td>1933</td>
<td>Farber et al.</td>
<td>3 days</td>
<td>Cyanosis and dyspnea since birth</td>
<td>No murmur</td>
<td>None</td>
<td>Thick in L.V.</td>
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<tr>
<td>13</td>
<td>1935</td>
<td>McIntosh et al.</td>
<td>34 mos</td>
<td>Bronchopneumonia at 21 mos.</td>
<td>Soft syst. murmur; card. failure</td>
<td>None</td>
<td>Thick plaques in L.A.</td>
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<tr>
<td>14</td>
<td>1938</td>
<td>Newns</td>
<td>21 mos</td>
<td>Anemia; dyspnea; no cyanosis</td>
<td>Loud M1; presyst. apical murmur; card. failure</td>
<td>None</td>
<td>—</td>
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<tr>
<td>15</td>
<td>1938</td>
<td>Fields</td>
<td>4 mos</td>
<td>Dyspnea; cyanosis</td>
<td>Loud M1; presyst. apical murmur</td>
<td>Aplasia left side of heart</td>
<td>—</td>
</tr>
<tr>
<td>16</td>
<td>1938</td>
<td>Fields</td>
<td>10 mos</td>
<td>—</td>
<td>Apical syst. murmur</td>
<td>P.D.A.; aplasia left side of heart</td>
<td>—</td>
</tr>
<tr>
<td>17</td>
<td>1938</td>
<td>Fields</td>
<td>4 days</td>
<td>Cyanosis</td>
<td>—</td>
<td>—</td>
<td>—</td>
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<tr>
<td>18</td>
<td>1938</td>
<td>Fields</td>
<td>3½ mos</td>
<td>Cyanosis; dyspnea</td>
<td>—</td>
<td>—</td>
<td>—</td>
</tr>
<tr>
<td>19</td>
<td>1941</td>
<td>Gross</td>
<td>4 days</td>
<td>Sudden cyanosis and dyspnea</td>
<td>—</td>
<td>—</td>
<td>P.D.A. — large; aort. sten.</td>
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### Table 4.—Continued

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<thead>
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<th>Case No.</th>
<th>Year</th>
<th>Reference</th>
<th>Age</th>
<th>History</th>
<th>Physical findings</th>
<th>Investigation</th>
<th>Pathology</th>
<th>Associated abnormality</th>
<th>Endocardium</th>
<th>Microscopic</th>
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<tbody>
<tr>
<td>20</td>
<td>1945</td>
<td>Johnson et al.</td>
<td>33 mos.</td>
<td>Pertussis and card. failure at 2½ yrs.</td>
<td>Syst. and pre-syst. murm at apex</td>
<td>—</td>
<td>Aort. sten.</td>
<td>Thick in L.A.</td>
<td>Fibrous scarring of endocard</td>
<td></td>
</tr>
<tr>
<td>21</td>
<td>1949</td>
<td>Craig</td>
<td>4 mos.</td>
<td>Feeding difficulty</td>
<td>No murmur</td>
<td>—</td>
<td>None</td>
<td>Thick in L.V.</td>
<td>Fibroelastic endocard; slight muscular degeneration</td>
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<tr>
<td>22</td>
<td>1949</td>
<td>Craig</td>
<td>7½ mos.</td>
<td>Feeding difficulty; cyanosis</td>
<td>Loud syst. and diast. murmur</td>
<td>—</td>
<td>Deformed tricuspid valve</td>
<td>Thick in L.A. and L.V.</td>
<td>Fibroelastic endocard; slight muscular degeneration</td>
<td></td>
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<tr>
<td>23</td>
<td>1949</td>
<td>Swan et al.</td>
<td>18 yrs.</td>
<td>Dyspnea; under-developed; card. failure; died during operation for coarc. &amp; P.D.A.</td>
<td>Syst. and diast. murmur in pulmon. area; accent. P$_2$; cyanotic toes</td>
<td>E.C.G: Prom. P waves; R.A.D; card. cath.</td>
<td>Coarc. of aorta; P.D.A. into desc. aorta</td>
<td>Normal</td>
<td>Severe pulm. vasc. changes</td>
<td></td>
</tr>
<tr>
<td>24</td>
<td>1950</td>
<td>McConnell</td>
<td>10 mos.</td>
<td>Well to 18 mos; weakness &amp; dyspnea; terminal cyanosis</td>
<td>Syst. murmur at apex &amp; rt. sternal border. card. failure</td>
<td>—</td>
<td>Tricuspid sten; aortic sten.</td>
<td>Thick</td>
<td>Fibrosis of myocardium; no inflammatory cells</td>
<td></td>
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<td>25</td>
<td>1951</td>
<td>Emery et al.</td>
<td>36 mos.</td>
<td>Well to 2 yrs. cough; dyspnea</td>
<td>No murmur; card. failure; heart not enl.</td>
<td>—</td>
<td>None</td>
<td>Pale in L.A.</td>
<td>Collagenous thick endocard; no inflammatory cells</td>
<td></td>
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<tr>
<td>26</td>
<td>1951</td>
<td>Emery et al.</td>
<td>18 mos.</td>
<td>Well to 17 mos. cough; bronchopneumonia and card. failure</td>
<td>Syst. and loud diast. murmur at apex; enl. heart only terminally</td>
<td>E.C.G: R.A.D.</td>
<td>Aortic valve, thickened edge; P.D.A. pinpoint</td>
<td>Thick in L.A.</td>
<td>Collagenous thick endocard; no inflammatory cells</td>
<td></td>
</tr>
<tr>
<td>27</td>
<td>1952</td>
<td>Blumberg et al.</td>
<td>Newborn</td>
<td>Cyanosis; respiratory distress</td>
<td>—</td>
<td>—</td>
<td>Aort. sten; P.D.A; P.F.O. —physiol.</td>
<td>Thick in L.A. and L.V.</td>
<td>Areas of degen. and calc. in myocardium</td>
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<td>No.</td>
<td>Authors</td>
<td>Duration</td>
<td>Symptoms</td>
<td>Signs of Left Heart Failure</td>
<td>Type of Lesion</td>
<td>Associated Lesions</td>
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<tr>
<td>28</td>
<td>Blumberg et al. 22</td>
<td>4½ mos.</td>
<td>Cyanosis since birth; card. failure</td>
<td>Loud syst. and diast. murmur at left sternal border</td>
<td>—</td>
<td>Aort. sten. P.D.A.</td>
<td>Thick in L.A. and L.V.</td>
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<td></td>
<td></td>
<td></td>
<td>Areas of degen. and fibrosis in myocardium</td>
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<tr>
<td>29</td>
<td>Blumberg et al. 22</td>
<td>16 mos.</td>
<td>Well to 15½ mos.; pneumonia; card. failure</td>
<td>No murmur</td>
<td>—</td>
<td>Thickening of aortic and pulm. valves</td>
<td>Thick in L.A. and L.V.</td>
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<td></td>
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<td></td>
<td></td>
<td>Fibroelastic tissue of endocard.</td>
<td></td>
<td></td>
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<tr>
<td>30</td>
<td>Blumberg et al. 22</td>
<td>16 mos.</td>
<td>Dyspnea and card. failure since 8 mos.</td>
<td>Syst. murmur at apex</td>
<td>—</td>
<td>None</td>
<td>Thick in L.A. and L.V.</td>
<td></td>
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<tr>
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<td></td>
<td>Fibroelastic tissue of endocard. invading myocardium</td>
<td></td>
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<tr>
<td>31</td>
<td>Azevedo et al. 23</td>
<td>18 mos.</td>
<td>Dyspnea &amp; fatigue; pneumonia; card. failure; died after ligation P.D.A.</td>
<td>Diast. rumble at apex; accent. split P; slight cyanosis of toes</td>
<td>E.C.G.: R.V.H. Prom. P waves; card. cath; angiocard.</td>
<td>P.D.A.</td>
<td>Marked pulm. vasc. changes</td>
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<td>32</td>
<td>Bower et al. 24</td>
<td>9½ mos.</td>
<td>Poor feeding; cough; card. failure; died after mitral valvotomy</td>
<td>Presyst. thrill and murmur; syst. murmur</td>
<td>E.C.G.: R.V.H. Prom. P waves; card. cath; angiocard.</td>
<td>None</td>
<td>Thick in L.A. and L.V.</td>
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<td>33</td>
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<td>None</td>
<td>None</td>
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<tr>
<td>34</td>
<td>Jacobson et al. 25</td>
<td>24 mos.</td>
<td>Dyspnea; poor development; died during operation for coarc. of aorta</td>
<td>Syst. murmur at rt. and left sternal borders; absent fem. pulsat.</td>
<td>E.C.G.: R.V.H. angiocardi.</td>
<td>Coarc. of aorta</td>
<td>Thick in R.V. and L.V.</td>
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</tbody>
</table>

Abbreviations: P.D.A. = patent ductus arteriosus; P.F.O. = patent foramen ovale; L.A. = left atrium; L.V. = left ventricle; R.V. = right ventricle; R.V.H. = right ventricular hypertrophy; R.A.D. = right axis deviation; — = no record.
also operated upon a 5½ year old girl who is still living.

Circulatory Arrangements Associated with Mitral Stenosis

Congenital mitral stenosis may cause severe hemodynamic alterations and the development of hypertensive pulmonary vascular changes which affect adversely the course and prognosis of otherwise more benign or correctable malformations.

Taussig has pointed out that, in congenital mitral stenosis, hypertrophy of the right side of the heart may occur in fetal life. If the pressure in the left atrium is elevated, the normal right-to-left shunt through the foramen ovale is reduced, and most of the blood returning from the placenta reaches the systemic circulation by passing through the right ventricle, pulmonary artery and, via the patent ductus, into the aorta. The malformations usually associated with mitral stenosis should not alter this fetal circulatory arrangement significantly. The blood flow through the left side of the heart thereby being reduced, areas of stenosis in the aortic or subaortic regions are to a great extent bypassed. Following birth, however, the presence of other lesions can modify the effect of mitral stenosis in different ways. While the combination of defects may be variable, essentially three different arrangements may occur (fig. 4):

1. Mitral stenosis occurs alone or in association with an obstructive lesion of the left side of the heart or the aorta (that is, aortic stenosis or coarctation of the aorta). The pulmonary vascular changes are apparently due to mitral stenosis. The pulmonary pressure is increased, the pulmonary flow is reduced, and there is no shunt. Case 5 falls into this group.

![Circulatory alterations in congenital mitral stenosis](image_url)

**Fig. 4.** Circulatory alterations in congenital mitral stenosis. See text. Heavy lines indicate areas of increased pressure. Size of vessels drawn to indicate amount of blood flow.
Resection of the coarctation in this instance removed the last of a series of obstructions while the major lesion remained.

2. Mitral stenosis is associated with a defect proximal to the mitral valve allowing a left-to-right shunt to occur, for example, interatrial septal defect. This shunt reduces the pressure in the left atrium and the usual effect of mitral stenosis upon the pulmonary vascular bed is therefore relieved. The pulmonary flow is increased, but, as the pulmonary vascular bed is essentially normal and can passively accommodate a large flow of blood, severe pressure changes do not occur. This arrangement is found in Lutembacher's syndrome. A similar effect has been produced surgically by shunting operations between pulmonary and systemic veins for the relief of acquired mitral stenosis.24

3. Mitral stenosis is associated with a defect distal to the mitral valve (patent ductus or interventricular septal defect) and a shunt can occur in either direction, depending upon the relative resistances in the systemic and pulmonary circulations. If, as in the case of a patent ductus, the shunt occurs in the usual fashion from aorta into pulmonary artery (figs. 4, 3A), the lungs are exposed not only to the back pressure due to mitral stenosis, but also to a large pulmonary blood flow under elevated pressure, and both of these factors would promote arterial thickening. The larger amount of blood having to pass through the narrowed mitral valve accentuates the effect of the stenosis. It may be hoped that ligation of the ductus may lead to improvement by reducing the amount of blood which must pass through these two areas of high resistance. If the pulmonary vascular changes are severe, the amount of blood which the lungs can accommodate is reduced and the shunt through the patent ductus may become reversed (figs. 4, 3B). When this occurs, the ductus acts as an escape mechanism and any further increase in pressure will lead to a larger amount of blood passing from pulmonary artery into aorta. At this time ligation of the ductus offers no improvement and is probably contraindicated. In case 1, reversal of flow through the ductus was only intermittent, and it was hoped that the pulmonary vascular changes might still be reversible. In case 8, a patient who died in a cyanotic attack, the circulatory arrangement may have been of a similar nature. In this instance, again, there were two lesions, the interventricular septal defect with over-riding aorta and mitral stenosis, additive in their effect in promoting pulmonary vascular changes. The narrowed pulmonary bed acted in a fashion similar to a pulmonary stenosis and prevented blood from entering the lungs. Thus, the ventricular septal defect acted as an escape mechanism and an excessive flow of venous blood was directed into the systemic circulation.

Discussion

An analysis of nine cases found in the records of The Children's Memorial Hospital and of 34 cases from the literature would indicate that congenital mitral stenosis is not so rare a lesion as it is at present considered to be. Our nine cases were encountered in the course of 2067 consecutive autopsies performed since 1934, among which were 210 cases of congenital heart disease, representing an incidence of 0.43 per cent in the total series and 4.3 per cent of cardiac malformations. Associated malformations of the heart and the great vessels were frequently present. Among the 43 cases under discussion, there were only eight instances of isolated mitral stenosis. An analysis of the associated lesions reveals a high incidence of malformations involving the aortic valve, the aorta and ductus arteriosus. There were only two cases of associated ventricular septal defect, and we have not yet discovered a case of mitral stenosis, undeniably of congenital origin, associated with a significant defect of the interatrial septum. This fact may lend support to the opinion that the mitral stenosis of Lutembacher's syndrome is acquired. Apparently, defects of the atrial, ventricular, and aortic septa rarely occur with mitral stenosis. The embryologic significance of this fact is a matter of conjecture. The earlier reports in the literature have been concerned with a controversy
regarding the occurrence of intrauterine inflammatory processes and their possible relationship to valvular malformations, but the theory of "fetal endomyocarditis" appears to have been disproved by later work based on careful microscopic study. Craig, in a study of cases of congenital heart disease with ventricular endocardial thickenings, pointed out the association of valvular lesions and the rarity of septal defects. He considered these lesions to be the result of a developmental abnormality consisting in an overproduction of tissues arising from the endothelial layer involving also, therefore, the closing masses of the various communications between the two sides of the heart. It is not the purpose of this paper to enter into a discussion of embryologic theories, but merely to point out that the selective association of certain malformations with congenital mitral stenosis may represent significant evidence for the future elucidation of the nature of these lesions.

Of the 43 cases under discussion, all but one died under 3 years of age. This finding testifies to the severe consequences of this malformation, for, in most instances, the associated defects (table 1) did not appear severe enough to be responsible for death at such an early age.

An analysis of the histories and physical findings has yielded no definite diagnostic criteria, but certain features of importance have become apparent. Eleven patients showed good health in early infancy and a sudden onset of symptoms, usually respiratory distress, at varying ages between 3 months and 2 years. This sudden onset of symptoms may be associated with the progressive hypertensive changes in the pulmonary vascular bed which, having reached a certain degree, precipitate the patient's rapid downhill course.

Cyanosis, often terminal, was observed in 19 instances and there were three cases of differential cyanosis involving the lower extremities due to shunting of blood from pulmonary artery to aorta through a patent ductus. Such reversal of blood flow may occur through a patent ductus present as an isolated malformation but complicated by pulmonary hypertension. As mitral stenosis is responsible for the development of severe pulmonary vascular changes and consequently a high pulmonary resistance, its presence should be considered when reversal of blood flow through a patent ductus is observed. From a consideration of the hemodynamic alterations imposed by mitral stenosis in the presence of defects distal to the mitral valve, it would appear that a significant degree of cyanosis in these cases is indicative of severe pulmonary vascular changes and a grave prognosis.

The auscultatory finding of an apical diastolic murmur, so characteristic of acquired mitral stenosis, was rarely present in these cases. Furthermore, the frequency of an apical diastolic rumble in congenital heart lesions in the absence of mitral stenosis is recognized, and the diagnosis of congenital mitral stenosis would be made too frequently if undue attention were directed toward this auscultatory sign. A more significant finding may be the accentuation of both the mitral first and the pulmonic second sounds, but, unfortunately, little attention has been paid to these signs in the histories. In six patients there were no murmurs.

In the electrocardiogram, a pattern of right ventricular hypertrophy was always demonstrated in the precordial leads. The P waves did not contribute helpful evidence of left atrial enlargement, nor was this abnormality satisfactorily detected on fluoroscopic examination. Cardiac catheterization in cases of acquired mitral stenosis usually demonstrates an elevated pulmonary "capillary" pressure. Such a finding may be of diagnostic value in cases of congenital heart disease where mitral stenosis is suspected, if left ventricular failure can be eliminated as its cause. The demonstration of delayed emptying of the left atrium by angiocardiography has provided an important indication of mitral stenosis.

From a consideration of these observations, it may be concluded that mitral stenosis should be suspected when the history is suggestive of pulmonary edema, when the clinical picture produced by recognized lesions, such as patent ductus or coarctation of the aorta, is atypical and when the electrocardiogram...
indicates a disproportionate degree of right ventricular hypertrophy.

Microscopic examination of the lungs of the nine cases encountered in this hospital has shown that extensive changes, similar to those found in acquired disease, can be present at an early age. The structural changes vary with the age of the patient and the degree of mitral stenosis. The severe narrowing of the small pulmonary arteries and arterioles observed in a child 2 years of age and illustrated in figure 3 is indicative of the progressive nature of these lesions. It is now well recognized that the presence of pulmonary hypertension and the anatomic alterations produced thereby are of great significance in determining the prognosis in the individual patient. This contention is supported by the early mortality in the group of cases under discussion and furthermore by the serious consequences of surgical correction of an associated lesion.

Surgical treatment of the congenital form of mitral stenosis has been attempted, but only one patient is known to have survived the procedure. A mitral valvulotomy, in cases of isolated mitral stenosis, or a valvulotomy combined with resection of a coarctation or ligation of a patent ductus is doubtless the desirable approach in the treatment of these patients. An evaluation of the structural alterations of the mitral valve observed in our cases suggests that, in congenital stenosis, the entire valve mechanism is affected and the excellent results which attend therapy of the acquired form may not be readily duplicated.

**Summary**

1. A clinicopathologic study of nine cases of congenital mitral stenosis is presented. This represents an incidence of 4.3 per cent among cases of cardiac malformations encountered in the autopsy records of this hospital.

2. A review of the literature since 1846 has revealed 34 cases.

3. Mitral stenosis occurred infrequently as an isolated malformation. Its association with aortic stenosis, coarctation of the aorta and patency of the ductus arteriosus is significant. Defects of the atrial, ventricular, or aortic septa were rarely present.

4. The diagnosis is difficult. The presence of mitral stenosis should be suspected when the history is suggestive of the occurrence of pulmonary edema, when the clinical picture produced by other lesions, such as coarctation of the aorta or patent ductus, is atypical, and when the electrocardiogram shows evidence of right ventricular hypertrophy in the presence of a recognized left-sided lesion. The finding of an elevated pulmonary “capillary” pressure on cardiac catheterization, and the demonstration of delayed emptying of the left atrium by angiocardiography are features of diagnostic importance.

5. Pathologic findings are noted. The appearance of the mitral valve was similar in all cases, showing thickened leaflets and short fused chordae tendineae, due to an irregular arrangement of cellular mesenchymatous tissue replacing the normal pattern of the endocardial layers. The right ventricle was hypertrophied in all instances. Pulmonary vascular changes were present in all but one case and were of a severe degree in the older patients.

6. Circulatory derangements imposed by mitral stenosis are discussed. If mitral stenosis occurs as an isolated lesion, its effect is similar to that of the acquired form. In association with aortic stenosis or coarctation of the aorta, it is one of a series of lesions obstructing the passage of oxygenated blood to the periphery. Its effect may be minimal if the obstruction it presents is relieved by a defect of the interatrial septum. When a patent ductus or an interventricular septal defect is associated, pulmonary hypertensive changes may alter the usual direction of the shunt.

7. The importance of congenital mitral stenosis is stressed because it may complicate a malformation otherwise amenable to surgical therapy and nullify the successful correction of the associated lesion.

**Sumario Español**

Nueve casos de estenosis mitral congénita se presentan con un repaso de 34 casos de la literatura. Una incidencia alta de malformaciones de la válvula aórtica, de la aorta y del ducto arterioso es aparente. Hallazgos clínicos y resultados de procesos investigativos se han
CONGENITAL MITRAL STENOSIS

repasado. Estenosis mitral es responsable por
severas alteraciones circulatorias y el desarrollo
de cambios hipertensión pulmonar vasculares.
La malformación tiene consecuencias serias.
Esta asociada con una mortalidad temprana y
convierte en inefectiva la corrección quirúrgica
de coartación de la aorta y del ducto arterioso
patente.

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Congenital Mitral Stenosis
CHARLOTTE FERENCZ, ARNOLD L. JOHNSON and F. W. WIGLESWORTH

Circulation. 1954;9:161-179
doi: 10.1161/01.CIR.9.2.161

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on
the World Wide Web at:
http://circ.ahajournals.org/content/9/2/161

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