Diseases of the Mitral Valve in Infancy

An Anatomic Analysis of 55 Cases

By Farzin Davachi, M.D., James H. Moller, M.D., and Jesse E. Edwards, M.D.

SUMMARY
A pathologic study of mitral valvular lesions among 55 infants revealed 29 cases in which the mitral valve was the site of a primary congenital malformation and 26 cases in which the mitral lesion was an acquired infarction of papillary muscles secondary to some other malformation.

Among the 29 cases with primary congenital anomalies of the mitral valve, there were 41 lesions represented. Congenital anomalies were identified according to the four components of the valve as follows: leaflets, commissures, chordae tendineae, and papillary muscles. The most common basis for primary congenital mitral valvular disturbance was an abnormality of the papillary muscles. In this group, parachute mitral valve and abnormal position of papillary muscles associated with endocardial fibroelastosis were most common (eight and 10 examples, respectively). Anomalous mitral arcade and obstructing papillary muscles were observed four and three times, respectively.

Involvement of leaflets was the second most common type of congenital anomaly, being observed 11 times. In this group, in order of decreasing frequency, were supravalvular ring (five cases), accessory mitral valvular tissue (three cases), "Ebstein's" malformation of the left atrioventricular valve in corrected transposition (two cases), and cleft mitral valve (one case).

Commissural fusion was observed once and, in two cases, involvement of multiple components of the valve was observed.

Among the 26 examples of infarction of papillary muscles, exclusive of 10 cases with endocardial fibroelastosis, the fundamental congenital anomalies included aortic stenosis (15 cases), coarctation of the aorta (six cases), and anomalous origin of the left coronary artery from the pulmonary trunk (five cases).

Additional Indexing Words:
Mitral stenosis Mitral insufficiency

In THE INFANT suffering from mitral stenosis or insufficiency, the anatomic cause may be either a primary anomaly of the valvular mechanism or an acquired state secondary to some other condition. Although many of the several anomalies have been reported individually, it seemed appropriate for us to view the entire material at our disposal so as to determine relative incidence and also to present a comprehensive picture of the subject. For this reason, an anatomic review was made of 55 specimens from subjects under the age of 2 years in whom mitral valvular disease either had been recognized clinically or was first identified by pathologic examination. In each case, the specimen was available for restudy. Fifty-four specimens were obtained at necropsy, and one was a mitral valve obtained at operation.

This study on the pathology of mitral valvular disease in the infant is presented.

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from the perspective of deviations from the normal in the four anatomic components of the mitral valve mechanism, namely, the leaflets, the commissures, the chordae tendineae, and the papillary muscles. In each case studied, the details of the mitral valve were considered according to the presence or absence of abnormality in the four named components of the valve. Associated conditions, when present, were identified.

Cases with mitral valvular disease in association with persistent common atroventricular canal, single ventricle, complete transposition of the great vessels, and those with asplenia or polysplenia were excluded from this study. Also, cor triatriatum was not included. Cases of congenitally corrected transposition of the great vessels exhibiting abnormalities of the left atroventricular valve were included. This selection yielded a total of 55 specimens for this study, among which were 67 abnormalities of mitral valvular components. As a point of reference, the essential anatomic aspects of the normal mitral valve are first presented.

**Structure of the Normal Mitral Valve**

The mitral valve is equipped with two leaflets, the anterior (septal) and the posterior (parietal); the two bridges of valvular tissue that connect the two leaflets are commonly called the commissures, the anterolateral and the posteromedial. The commissures correspond with subjacent groups of chordae and papillary muscles (fig. 1a). Chordae tendineae run between each leaflet to the anterolateral and posteromedial papillary muscles of the left ventricle. Conversely, from each papillary muscle a set of chordae runs to the corresponding aspect of each leaflet.

On the basis of these anatomic arrangements, during ventricular diastole, flow of the blood into the left ventricle may pass through two channels, the primary and secondary orifices of the mitral valve (fig. 1b). The primary orifice is bounded anteriorly and posteriorly by the two leaflets and laterally and medially by the two papillary muscles. The secondary orifices are a series of openings lying between the chordae tendineae. Flow through these orifices allows blood to enter the left ventricle lateral to one papillary muscle and medial to the other.

During ventricular systole, closure of the mitral valve is accomplished by apposition of its two leaflets. It is evident that, to form a complete partition between the atrium and the ventricle,
## Table 1

### Summary of 41 Primary Anomalies of the Mitral Valve in 29 Infants

<table>
<thead>
<tr>
<th>Anatomic site</th>
<th>No. of cases</th>
<th>Case no. in subgroup</th>
<th>Associated mitral anomalies</th>
<th>Associated anomalies other than mitral</th>
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<tbody>
<tr>
<td>Leaflets</td>
<td></td>
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<td></td>
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<tr>
<td>Supravalvular ring</td>
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<td>1</td>
<td>Mild commissural fusion; accessory tissue, mitral valve</td>
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<tr>
<td></td>
<td></td>
<td>2</td>
<td>Parachute mitral</td>
<td>+ + 0 OBGVRV</td>
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<tr>
<td></td>
<td></td>
<td>3</td>
<td>Parachute mitral</td>
<td>+ 0 + -</td>
</tr>
<tr>
<td></td>
<td></td>
<td>4</td>
<td></td>
<td>0 0 0 OBGVRV</td>
</tr>
<tr>
<td></td>
<td></td>
<td>5</td>
<td></td>
<td>0 + + -</td>
</tr>
<tr>
<td>Cleft mitral</td>
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<td>1</td>
<td>0</td>
<td>+ 0 0 Tricuspid pouch</td>
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<tr>
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<td>3</td>
<td>1</td>
<td>0</td>
<td>0 0 + Hypoplasia of LV, EFE</td>
</tr>
<tr>
<td></td>
<td></td>
<td>2</td>
<td>Supravalvular ring LA</td>
<td>+ 0 + -</td>
</tr>
<tr>
<td></td>
<td></td>
<td>3</td>
<td>Parachute mitral</td>
<td>0 0 0 -</td>
</tr>
<tr>
<td>“Ebstein’s”</td>
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<td>1</td>
<td>0</td>
<td>+ 0 0 -</td>
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<tr>
<td></td>
<td></td>
<td>2</td>
<td>0</td>
<td>+ 0 0 -</td>
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<tr>
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<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fusion</td>
<td>1</td>
<td>1</td>
<td>Supravalvular ring</td>
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<tr>
<td>Chordae tendineae</td>
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<td></td>
<td></td>
<td></td>
</tr>
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<td>Short</td>
<td>2</td>
<td>1–2</td>
<td>Multiple of mitral valve</td>
<td>-</td>
</tr>
<tr>
<td>Papillary muscles</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Anomalous arcade</td>
<td>4</td>
<td>1</td>
<td>0</td>
<td>0 0 0 Mild AS</td>
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<tr>
<td></td>
<td></td>
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<td>0</td>
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</tr>
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<td></td>
<td>3</td>
<td>0</td>
<td>0 0 0 -</td>
</tr>
<tr>
<td></td>
<td></td>
<td>4</td>
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<tr>
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<td>1–8</td>
<td>See table 2</td>
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<tr>
<td>Abnormal position</td>
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<td>1–10</td>
<td>0</td>
<td>0 0 0 Each EFE</td>
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<tr>
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<td>1</td>
<td>Mitral arcade</td>
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</tr>
<tr>
<td></td>
<td></td>
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<td>0 0 0 ASD</td>
</tr>
<tr>
<td>Multiple components of mitral valve</td>
<td>2</td>
<td>1</td>
<td>-</td>
<td>+ 0 0 OBGVRV</td>
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<tr>
<td></td>
<td></td>
<td>2</td>
<td>-</td>
<td>+ 0 0 -</td>
</tr>
</tbody>
</table>

Abbreviations: OBGVRV = origin of both great vessels from right ventricle; EFE = endocardial fibroelastosis; AS = aortic stenosis; LV = left ventricle; LA = left atrium; PDA = patent ductus arteriosus; VSD = ventricular septal defect.
the presence of an adequate amount of the valvular tissue is essential and the leaflets must be able to move into appropriate positions. Free motion of the mitral leaflets depends on three factors: (1) the presence of pliable valvular tissue, (2) lack of any undue union or restraint at the commissural junctions, and (3) proper position, function, and length of the chordae and papillary muscles. Chordae and papillary muscles exert a certain restraint upon the leaflets so that at the height of systole, the apposition of the leaflets is arrested at an optimal level for anatomic closure of the valve.

Observations

Primary Anomalies of the Mitral Valve (table 1)

Among the material studied, the cases fell into two groups, one in which the mitral valvular disease was primary and congenital in nature and the other group in which the mitral valvular disease was acquired secondary to an anomaly involving some other structure of the heart. This section will consider the first group.

Anomalies Involving the Leaflets

A number of anomalies may involve the mitral leaflets as to cause valvular dysfunction. These will be considered in sequence.

Supravalvular Ring of the Left Atrium. In this entity, a circumferential ridge of connective tissue is attached to the base of the atrial surfaces of the mitral leaflets and protrudes into the orifice of the mitral valve. Depending upon the diameter of the ring, various degrees of obstruction to flow into the mitral valve result. In most cases, no significant obstruction occurs but in the relatively few cases with the fully developed deformity, the supravalvular ring acts as a significantly stenosing diaphragm.

In five of the 55 cases in this review, a supravalvular ring of the left atrium was observed. The ages of the patients ranged from 2 to 9 months. There was one male and four females. In two cases, ages 3 and 9 months, respectively, significant obstruction existed, while in the remaining three cases no significant obstruction was present. Obstruction was associated with enlargement of the left atrium and marked thickening of its endocardium.

In the case with the greatest degree of obstruction, that of the 9-month-old infant, the diameter of the orifice of the supravalvular ring was only 3 mm (fig. 2). Jet lesions were present upon the septal aspect of the left ventricle opposite the site of the severe stenosis of the supravalvular ring. Associated anomalies in this case were (1) mild fusion of the commissures of the mitral valve, (2) coarctation of the aorta, (3) ventricular septal defect, and (4) accessory tissue of the mitral valve. Associated anomalies in the other case with a significant degree of obstruction at the ring were (1) origin of both great vessels from the right ventricle, (2) ventricular septal defect, (3) parachute deformity of the mitral valve, (4) subaortic stenosis, and (5) anomalous attachment of the chordae tendineae to the rim of the ventricular septal defect. The latter process obstructed the ventricular septal defect. In one of the three cases without obstruction at the ring, associated anomalies were parachute deformity of mitral valve, coarctation of the aorta, and ventricular septal defect. Another case exhibited origin of both great vessels from the right ventricle. In the final case, coarctation of the aorta and subaortic stenosis were associated.

Cleft Mitral Leaflet. A cleft in the anterior leaflet of the mitral valve is commonly present in classic examples of persistent common atrioventricular canal, but this condition may occur infrequently as an isolated condition. In our series, this anomaly was observed in one case involving a 4-month-old female infant. No distinct pathologic evidence of mitral insufficiency was identified (fig. 3). Associated anomalies in this case were ventricular septal defect and anomalous attachment of chordae between the edges of the valvular cleft, on one hand, and the rim of the ventricular septal defect, on the other. An interesting phenomenon was the presence of redundant valvular tissue involving the tricuspid valve. This formed a pouch which lay in a position capable of closing the ventricular septal defect.
Figure 2

Supravalvular ring of the left atrium; case of 9-month-old infant. (a) Superior view of left atrium (L. A.). Arrows indicate the markedly narrowed orifice of the stenosing ring. Atrial septal defect of fossa ovalis type (A. S.). Left atrial endocardium is markedly thickened. (b) Supravalvular ring. Mitral valve, left atrium (L. A.), and left ventricle (L. V.) have been opened. Arrows indicate the supravalvular ring. (c) Low power photomicrograph through the supravalvular ring (R.). Left atrium (L. A.) above, the left ventricle (L. V.) below. P.M. = posterior mitral leaflet. (From Rao and associates;26 with permission.)
Cleft mitral leaflet; from a 4-month-old female in whom a ventricular septal defect was also present. Left atrium (L. A.) above, left ventricle (L. V.) below. The cleft (between arrows) involves the center of the anterior leaflet. A few short chordae tendineae extend between base of cleft and the ventricular septum.

Accessory Orifice of Mitral Valve. The condition called accessory orifice of the mitral valve is characterized by a variably-sized circular deficiency in the substance of a valve leaflet. From the circumference of the accessory orifice, chordae tendineae pass to be inserted into an independent papillary muscle. The valve may be incompetent. No examples of this condition were in this series.

Accessory Mitral Valvular Tissue. Three examples of accessory mitral valvular tissue were encountered. One of the subjects was male and two were female. The ages ranged from 10 weeks to 9 months. In each case, a large accessory papillary mass was attached to the atrial aspect of the posterior leaflet of the mitral valve (fig. 4). On the basis of its size, the accessory tissue was capable of obstructing the orifice of the valve. In one case, involving a 7-month-old infant with coarctation of the aorta, hypoplasia of the left ventricle and endocardial fibroelastosis were associated, each of which seemed to have contributed, along with the accessory tissue, to evident pulmonary venous obstruction. In the other two cases, one with supravalvular ring of the left atrium and the other with parachute deformity of the mitral valve, clinically observed signs of mitral valvular obstruction were present.

"Ebstein's" Malformation of Inverted Tricuspid Valve. In congenitally corrected transposition of the great vessels, the ventricles and atroventricular valves are inverted, so that the left atroventricular valve has the structure of a tricuspid valve. The inverted tricuspid valve, then, is subject to Ebstein's malformation as is the case with the normally situated tricuspid valve. We observed two male infants, one a stillborn and the other 7½ months old, with congenitally corrected transposition of the great vessels and coexistent Ebstein's malformation of the inverted tricuspid valve. In each case, a ventricular septal...
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A 9-month-old female infant. The anatomic appearance of the mitral valve in this case was similar to that observed in mitral stenosis of rheumatic origin. There was fusion of both commissures resulting in a funnel-shaped valve. Associated lesions in this case were supravalvular ring of the left atrium and ventricular septal defect.

Anomalies Involving the Chordae Tendineae

Abnormal insertion or length of the chordae tendineae may interfere with the functional integrity of the mitral valve. The cases described in this section represent isolated anomalies of the chordae tendineae.

**Short Chordae Tendineae.** Short chordae tendineae limit mobility of the mitral valve leaflets. The leaflets, therefore, become fixed along the wall of the left ventricle and are unable to contribute to closure of the mitral valve. Mitral insufficiency consequently develops. In this series, we did not observe cases in which short chordae tendineae represented an anomaly.

**Anomalies Involving the Commissures**

**Congenital Commissural Fusion.** Commissural fusion was observed in one case, that of a stillborn infant with corrected transposition of great vessels. Opened left atrium (L. A.) and left-sided ventricle ("L. V."). Left side of heart. The origin of the inverted tricuspid valve (solid line) is below the normal position of the annulus (dotted line). Chordae are short and attached to nonspecialized portions of ventricle. The left atrium is markedly enlarged. Marked endocardial thickening of ventricle.

"Ebstein's" anomaly of inverted tricuspid valve in a stillborn infant with corrected transposition of great vessels. Opened left atrium (L. A.) and left-sided ventricle ("L. V."). Left side of heart. The origin of the inverted tricuspid valve (solid line) is below the normal position of the annulus (dotted line). Chordae are short and attached to nonspecialized portions of ventricle. The left atrium is markedly enlarged. Marked endocardial thickening of ventricle.

Defect was present. The left atrioventricular valve manifested downward displacement of the posterior and septal leaflets characteristically present in Ebstein's malformation of the tricuspid valve (fig. 5). The cavity of the left-sided (arterial) ventricle was somewhat small as a result of the low attachment of the malformed leaflets. The left atrium was enlarged secondary to "mitral" regurgitation.

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isolated malformation. We have, however, observed this condition in two cases with involvement of several elements of the mitral valve mechanism. These will be described in the section dealing with anomalies involving more than one component of the mitral valve.

Long Chordae Tendineae. Among the 55 cases of this study, which were limited to patients under 2 years of age, there were no examples in which long chordae tendineae were a basis for mitral valvular dysfunction. In our pathologic collection, there is the case of a child, 2 years and 10 months old, with this condition and Marfan's syndrome, a case reported by Raghib and associates. Although involvement of the aorta in Marfan's syndrome is well recognized, severe involvement of the mitral valve in infancy is uncommon. In this child, the chordae tendineae were greatly elongated, so that during ventricular systole the leaflets did not come into apposition but ballooned into the left atrium resulting in mitral regurgitation. This process may have been accentuated by coexistent large redundant mitral leaflets. A greatly enlarged left atrium and left ventricle were evidence that marked mitral insufficiency had been present in this case.

Anomalies Involving the Papillary Muscles

Anomalous Mitral Arcade. In anomalous mitral arcade, the anterolateral and posteromedial papillary muscles of the left ventricle and anterior mitral leaflet together form an arcade. In this condition, described by Layman and Edwards, a bridge of fibrous tissue which is continuous with the free aspect of the anterior mitral leaflet arches between the two papillary muscles (fig. 6). The mitral commissures are not fully developed. The chordae tendineae are somewhat thickened and short, so that the papillary muscles are essentially in direct continuity with the anterior mitral leaflet. Histologic examination of the arcade reveals a composition of connective tissue with areas of dense collagenous material.

Anomalous mitral arcade was present in four infants, ranging in age from 2 weeks to 11 months, one of whom is a living patient from whom the surgically excised specimen of mitral valve was available for study. In one of the three necropsied patients, mild aortic stenosis was coexistent. Each of these specimens revealed evidence of mitral insufficiency. The fourth example of mitral arcade was present in the surgically removed mitral valve mentioned. It was from an 11-month-old infant with signs of mitral valvular obstruction. In addition to an anomalous mitral arcade, the specimen revealed greatly hypertrophied papillary muscles which resulted in obstruction at a level below the mitral valve.

Parachute Mitral Valve. The developmental complex of parachute mitral valve and associated obstructive lesions in the left side of the heart is now a recognized entity. The comprehensive first report of this condition by Shone and associates described the parachute mitral valve in eight patients, the youngest being 2 years and 9 months old. Since that time, we observed eight additional specimens with parachute mitral valve in patients under 2 years of age. Four of these patients were male, and four were female.

In parachute mitral valve, a single left ventricular papillary muscle is present into which the chordae tendineae from both mitral leaflets insert (fig. 7). The valvular leaflets and commissures are normal. Mitral stenosis may be associated with parachute mitral valve because of several anatomic features. The chordae tendineae in this condition are usually short and thickened, thereby allowing only limited movement of the leaflets. The primary orifice of the mitral valve is reduced in size as a result of the convergence of all the chordae tendineae into the solitary papillary muscle, so that blood flow occurs through the secondary orifices, between the chordae. The secondary orifices, however, are frequently obstructed by the thickened and fused chordae.

Although only one papillary muscle is present in the classical form of parachute deformity of the mitral valve, we have observed a case in which two papillary muscles were present but so close together as...
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Figure 7
Parachute mitral valve. (a.) Case 3 of table 2. Left side of heart. The chordae tendineae from both mitral valvular leaflets are inserted into a single papillary muscle (P.), forming a funnel-shaped mitral valve. Chordae tendineae are short and fused with narrow interchordal spaces. (b.) Case 1 of table 2. Variant of parachute mitral valve. Left ventricle and aortic valve (A. V.). Although two left ventricular papillary muscles (P.) are present, these lie so close to each other as to yield a variant of the parachute mitral valve.

Abnormal Position of Papillary Muscles.
In endocardial fibroelastosis, the papillary muscles arise high on the left ventricular wall.11 Contrasted to the normal heart wherein the papillary muscles arise at the junction of the middle and lower thirds of the left ventricle, in this condition they arise from the upper third of the ventricular wall. The papillary muscles are also smaller than normal and may reveal histologic changes of infarction. From the apices of the papillary muscles, short, thickened chordae pass in a lateral direction to be inserted into the mitral valvular leaflets. As a result, the mitral valve is tethered and regurgitant. Anatomic changes to border on being a single muscle. In this case, the chordae tendineae converged to be inserted into the two closely related papillary muscles. Clinical signs of mitral stenosis had been present in the patient.

Among the eight new cases with parachute mitral valve, evidence for significant obstruction was present in seven. The associated anomalies observed in our eight cases are listed in table 2. This shows that the most commonly associated condition was left ventricular outflow obstruction. Coarctation of the aorta was less common. Origin of both great vessels from the right ventricle was observed in three of the cases.
Table 2
Coexistent cardiac anomalies in eight infants with parachute mitral valve (cases other than those reported by Shone10). A. S. or sub. A. S. = aortic or subaortic stenosis; V. S. D. = ventricular septal defect; O. B. G. V. from R. V. = origin of both great vessels from right ventricle; P. S. = pulmonary stenosis; A. S. D. = atrial septal defect.

<table>
<thead>
<tr>
<th>Case No</th>
<th>Sex</th>
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<th>Parachute Mitral</th>
<th>A. S. or Sub A. S.</th>
<th>Coarct of Aorta</th>
<th>Supravalvular Ring</th>
<th>Associated Lesion</th>
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<td>1</td>
<td>M</td>
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<td></td>
<td>V.S.D.</td>
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<td>2</td>
<td>F</td>
<td>3</td>
<td></td>
<td></td>
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</tr>
<tr>
<td>3</td>
<td>F</td>
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<tr>
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<td>F</td>
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<td></td>
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</tbody>
</table>

- Significant obstruction
- Obstruction present, but not significant
- Not present

Coexistence of parachute mitral valve and subvalvular stenosis is a feature of parachute mitral valve syndrome. Significant obstruction is present in all cases, but is not obstructive in all cases.

Obstruction by Abnormal Papillary Muscles. Three specimens were observed in which the mitral valve was obstructed by subjacent large papillary muscles. Although the leaflets of the mitral valve were normal, the bulky papillary muscles, which were situated immediately below the valvular orifice, obstructed the flow of blood from the left atrium to the left ventricle (fig. 8). Two hearts and one surgical specimen were available for this study from two male infants and one female infant, whose ages ranged from 2 days to 11 months. The surgical specimen represented resection of the mitral valve and its papillary muscles from an 11-month-old male infant (fig. 9). In this specimen, an anomalous mitral arcade was also present. Reference to this case was made in a previous section. With the placement of a prosthetic valve, clinical improvement of the infant's condition resulted.

Among the two necropsy specimens with obstructing papillary muscles, one case showed an associated patent ductus arteriosus representing regurgitation are observed in most cases. We observed these anatomic changes in ten infants, each with endocardial fibroelastosis, whose ages ranged from 3 hours to 2 years.

Obstruction of mitral orifice by abnormal papillary muscles. Case of 3-month-old infant with coexistent patent ductus arteriosus. Opened mitral valve. The papillary muscles are large and their apical ends lie so close to the valve orifice as to be obstructive. (a.) Posteromedial (P-M) papillary muscle. (b.) Anterolateral (A-L) papillary muscle.

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the coexistent ventricular septal defect was partially obstructed.

In the second case, a stillborn female infant, the posteromedial papillary muscle was hypoplastic (fig. 10b) and the chordae tendineae were short. The chordae were inserted directly into the free wall of the left ventricle, restricting the free motion of the valve leaflets, although evidence of mitral valvular malfunc-

and one an atrial septal defect at the fossa ovalis.

Anomalies Involving More than One Component of the Mitral Valve Mechanism

In two cases, complex abnormalities of the mitral valve mechanism were present. In a 3-month-old male infant with origin of both great vessels from the right ventricle, the mitral valve was slightly stenotic because of several features. Both papillary muscles were poorly developed and small (fig. 10a). The chordae tendineae were short, and several were attached directly to the ventricular septal wall. As a result of the anomalous attachment of the chordae to the septal wall,

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Figure 9
Obstruction by abnormal papillary muscles. Surgically removed specimen from an 11-month-old male infant. (From Castaneda and associates;12 with permission.) (a.) Mitral valve viewed from above. Ventricular orifice reduced in size by enlarged papillary muscles (P.). (b.) Lateral view of enlarged papillary muscles which fuse directly into mitral valve leaflets without distinct chordae being present.

Figure 10
Complex anomalies of mitral valve. (a.) Involvement of more than one component of the mitral valve in a case of a 3-month-old male infant. Opened left atrium (L. A.) and left ventricle (L. V.). Indistinct development of papillary muscles. Chordae tendineae are short and some are abnormally attached directly to the ventricular wall. (b.) Opened left atrium (L. A.) and left ventricle (L. V.) From a stillborn infant. Only the anterolateral set of papillary muscles (A-L) are formed. Units of this structure are enlarged and attached to short crossing chordae.
Infarction of papillary muscles in severe congenital aortic valvular stenosis; from a 7-month-old female infant. (a.) Thinning of posteromedial papillary muscle (P-M) associated with short chordae tendineae. (b.) Photomicrograph of papillary muscle and adjacent left ventricular wall. Fibrous tissue replacement of myocardium and interstitial cellular infiltration. The picture is that of healing myocardial infarction. Hematoxylin-eosin stain × 40.

Acquired Diseases of the Mitral Valve (Infarction of Papillary Muscles)

In several conditions, such as aortic stenosis, anomalous origin of a coronary artery from the pulmonary trunk, and endocardial fibroelastosis, the left ventricular papillary muscles may become infarcted, resulting in papillary muscle dysfunction and clinical evidence of mitral valvular insufficiency. One or both papillary muscles may be involved with the extent of the infarction varying considerably (fig. 11). Infarcted papillary muscles appear thin and flattened and may be discolored along the entire body or only toward their apices.

In some cases, the histologic appearance of the papillary muscles reveals acute myocardial infarction but, more frequently, the myocardium of the papillary muscle is replaced by various amounts of scar tissue as evidence of healed infarction. In some instances, calcification of the infarcted tissue is observed. The subendocardial muscle of the left ventricular wall may also show infarctive changes.

Evidence for mitral insufficiency is generally present, with left atrial enlargement being common. The free edge of the anterior leaflet of the mitral valve may be rolled and thickened, as a consequence of mitral regurgitation. In a few cases, “jet lesions” are observed on the posterior wall of the left atrium. In our series, there were examples of infarction of the papillary muscles and consequent development of the papillary muscle dysfunction with mitral insufficiency in association with one of the following conditions: aortic stenosis (15
cases), coarctation of the aorta (six cases), and anomalous origin of left coronary artery from the pulmonary trunk (five cases). In addition, there were ten examples of endocardial fibroelastosis with various degrees of infarction of papillary muscles. Each of the latter ten cases was considered in the section dealing with abnormal position of the papillary muscles.

Comment

Among 55 infants with mitral valvular lesions, 67 conditions were observed. This study showed that mitral valvular disorders responsible for symptoms in infancy present a more complicated clinical and pathologic picture than those involving children or adults. In infants, disorders of the mitral valve are more frequently associated with other cardiac malformations than they are in older subjects, yielding corresponding complicated manifestations.\(^{16,17}\) Also, unless special studies are performed, it may be difficult to determine whether known mitral valvular disease takes the form of stenosis or insufficiency.

In this review of our pathologic experience, we considered disorders on the basis of involvement of the four components of the mitral valve (leaflets, chordae tendineae, commissures, and papillary muscles). This approach permits insight into the function of each element of the valve and allows logical separation of cases into anatomic groups.

The occurrence of isolated mitral stenosis in the pediatric age group is uncommon. This pathologic state may result from several anatomic conditions, depending on which elements of the mitral valve are abnormal. In the study reported, the parachute mitral valve was the most frequent type of mitral stenosis. In this condition, the leaflets and commissures are normal. The mobility of the valve is limited, and the principal orifice is restricted from opening freely. The secondary orifices play an important role in flow of the blood from the left atrium to the left ventricle and, thus, the degree of the mitral stenosis depends upon the collective caliber of the openings of the interchordal spaces. Since, in the para-

chute mitral valve one cannot relieve the stenosis by opening the principal orifice, enlargement of interchordal spaces by selective resection of chordae may prove satisfactory. We are aware of one case thus operated with long range improvement, the patient being under the care of Drs. Harold Katkov and Frank E. Johnson of Minneapolis. Parachute mitral valve is almost uniformly associated with other cardiovascular malformations, particularly supravalvular mitral ring, subaortic stenosis, and coarctation of the aorta.\(^{10}\)

Supravalvular mitral ring may result in the findings of mitral valvular obstruction if the ring is extensive. The frequent association of this condition with parachute mitral valve is well recognized.

Mitrval obstruction may also result from abnormally large and malpositioned papillary muscles, as occurred in three of our cases. These unduly large papillary muscles arose at a site higher in the left ventricle than the normal sites and, by crowding the principal orifice of the mitral valve, caused obstruction to the egress of blood from the left atrium. This malformation was recognized in one of our cases during operation, and the entire valve and papillary muscles were resected and replaced by a valvular prosthesis.\(^{12}\)

A fourth anatomic cause of mitral stenosis is congenital commissural fusion. In this regard, these changes are similar to the basic alteration observed in rheumatic mitral stenosis. The leaflets of the mitral valve are thickened and fused at the commissures. In our one case, the chordae tendineae were short and inserted into poorly developed papillary muscles. The commissural fusion resulted in a funnel shape to the mitral valve, leading to an effective diminution of the principal and secondary orifices.

While deficiency in the substance of a leaflet may cause insufficiency, accessory mitral valvular tissue may become obstructive.\(^{18}\) We observed three cases, each showing an accessory fragment of valvular tissue attached to the posterior leaflet of the mitral valve. It may be worthwhile mentioning that coexistent endocardial fibroelastosis with hy-
poplastic left ventricle was present in one case.

Mitral insufficiency may result from one of several anatomic malformations. Deficiency in valvular tissue as a cause of mitral insufficiency is well understood. The anomaly may be represented by a void in substance involving either the free aspect of the leaflet as a cleft or the substance of a leaflet as a perforation. Of interest is the anatomic state associated with cleft mitral valve in which there are abnormal accessory chordae tendineae arising from the cleft or the central portion of the leaflet. These accessory chordae tendineae may exert an undue restraint upon the leaflet and, hence, exaggerate the insufficiency caused by the cleft.

A newly recognized cause of mitral insufficiency is anomalous mitral arcade. This is characterized by abnormal attachment of the papillary muscles to the leaflets with the formation of a fibrous arcade along the anterior aspect of the mitral leaflet. The principal orifice is normal or somewhat dilated, while the secondary orifices are diminished in caliber. Absence of the chordae or short chordae between the anterior mitral leaflet and the papillary muscles presents an undue restraint upon the mobility of the leaflet, preventing normal closure of the valve during ventricular systole.

Chordae tendineae play an important role in maintaining the integrity of the mitral valve. Their length and thickness, as well as their site of insertion, are important factors. Short chordae are often thick and, when those inserting into the posterior mitral leaflet are involved, apposition of the two leaflets becomes impaired on the basis of immobility of the leaflet.

While in Marfan's syndrome cystic medial necrosis of the aorta is a well recognized condition, the mitral valve may be involved as well. We have observed a young child, not in this series, in whom unusually long chordae tendineae appeared to be the basis for demonstrated mitral insufficiency.

Loss of the function of the papillary muscles results in loss of the optimal restraint upon the valve leaflets. This condition is commonly seen in infarction of the papillary muscles. Severe congenital aortic stenosis, coartation of the aorta, and anomalous origin of a coronary artery from the pulmonary trunk are the three conditions that cause infarction of the papillary muscles in infancy and lead to development of mitral insufficiency. In endocardial fibroelastosis, mitral insufficiency is common and may result from a combination of factors including left ventricular enlargement, peculiar mural insertion of papillary muscles, and infarction of the papillary muscles.

Although cases with complete transposition of the great vessels were not included in this study, it is recognized from an earlier study that mitral valvular anomalies are not uncommon in that condition. Many of the types seen in this study may be observed among patients with complete transposition. In the latter group, an observed condition of particular interest is abnormal attachment of the anterior mitral leaflet to the ventricular septum causing left ventricular outflow (subpulmonary) stenosis.

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