

بسم الله الرحمن الرحيم
الحمد لله الذي هدانا لهذا
ما كنا لنهتدي لولا أن هدانا الله

بسم الله الرحمن الرحيم

[قالوا سبحانك لا علم لنا إلا ما علمتنا إنك أنت العليم الحكيم]

صدق الله العظيم

[سورة البقرة - الآية ٢٢]

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CONGENITAL MITRAL VALVE ANOMALIES

REVIEW OF LITERATURE

SUBMITTED IN PARTIAL FULFILMENT OF

M. SC. DEGREE IN CARDIOLOGY

BY

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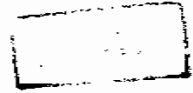
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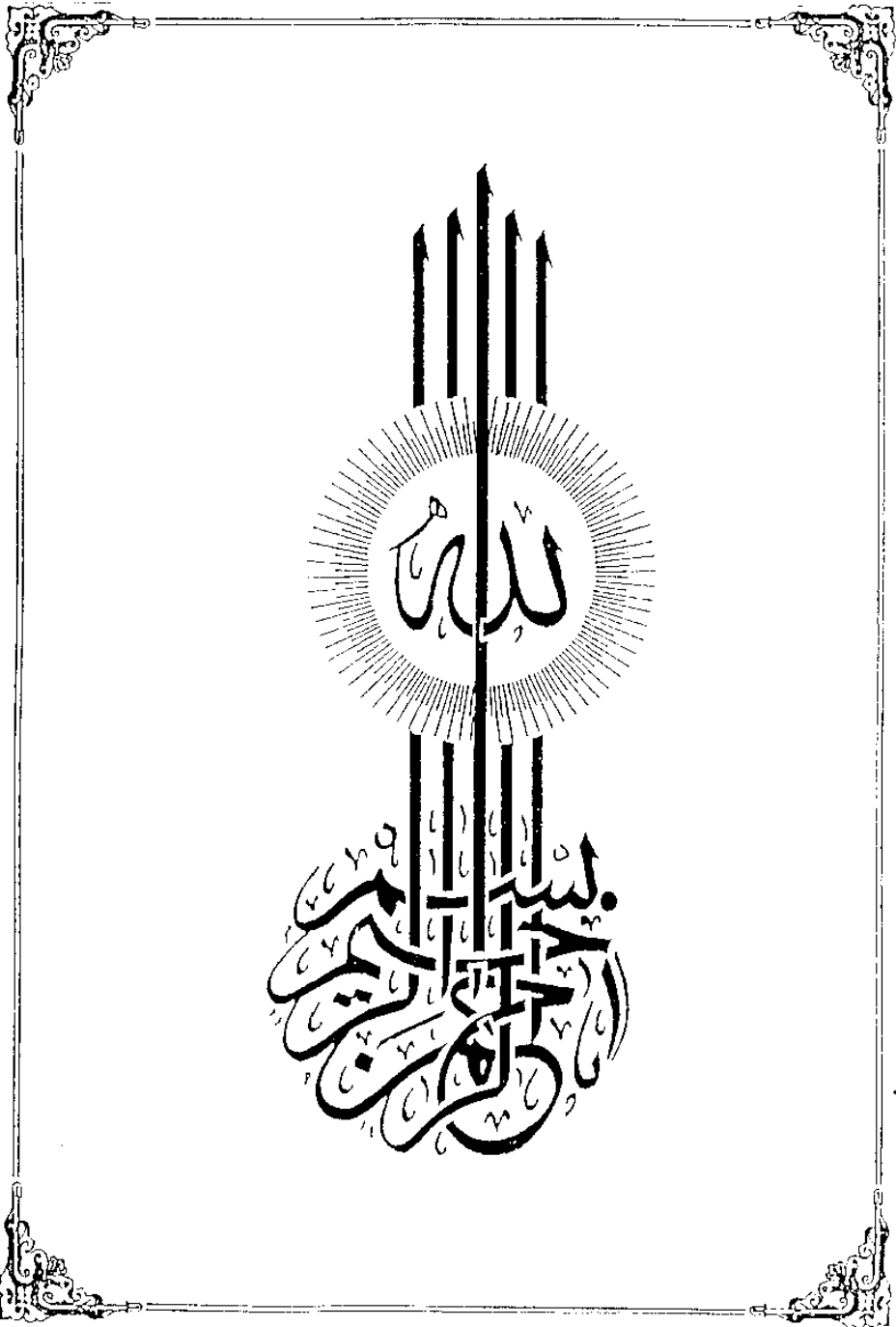
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INTRODUCTION

root. The left trigone is composed of fibrous tissue at the confluence of the left margin of the aortic and mitral valves. Between the trigones ventrally, the anterior mitral leaflet is in direct fibrous continuity with the aortic root (Ormiston JA, et al, 1981).

The mitral ring is usually incomplete anteriorly along the entire breadth of the dorsal attachment of the anterior mitral cusp (Walmsley R, 1978).

The leaflets

Anatomically they are the anterior (aortic) leaflet, the posterior (mural) leaflet, the lateral and the medial leaflets. (Kalmanson D, 1976). Physiologically they are the anterior and the posterior leaflets (Roberts WC, 1983).

The width of the basal attachment of the anterior leaflet is less than that of the posterior leaflet. (Di Segni E, and Edwards JE 1983), and the length of the annular attachment of the posterior leaflet is about twice that of the anterior leaflet (Roberts WC, 1983).

The anterior leaflet is more mobile, and the posterior leaflet has mainly a supporting role. (Perloff JK, and Roberts WC, 1972).

The anterior leaflet is semicircular or triangular in shape, on its atrial surface there is a ridge 0.8-1 cm from its free margin which delineate the line of leaflet closure, and its distal one third is rough, opaque, receives the insertion of the chordae tendineae on its ventricular surface (the rough zone) , while the rest of the leaflet is clear and membranous.

During valve closure the rough zone comes into apposition with its related part on the posterior leaflet. (Kalmanson D, 1976).

The anterior leaflet is one of the boundaries of the left ventricular outflow tract, and it has a common attachment with the left coronary cusp and half of the non coronary cusp of the aortic valve to the cardiac skeleton. (Ranganathan N, et al, 1970) , the ascending aorta, the membranous ventricular septum and the atrial septum (Roberts WC , 1983).

The posterior leaflet has a greater attachment to the mitral annulus than the anterior leaflet.(Kalmanson D, 1976). Its free margin possesses a number of clefts that divide it into three scallops, a big middle and two equal commissural scallops, one on each side (Ranganathan N, et al, 1970).

Three zones could be identified on the posterior leaflet surface:-

- a- A rough distal zone, has chordal insertion on its ventricular surface, it is opaque and comes into close contact with the anterior leaflet during valve closure.
- b- A clear zone proximal to the rough zone .
- c- A basal zone between the clear zone and the annulus, it receives the insertion of the basal chordae tendineae, which originate directly from the trabeculum carneae of the left ventricular myocardium (Lam JHC, et al, 1970).

The medial and lateral leaflets are usually bilobed

The chordae tendineae

From each papillary muscle a set of chordae run to the corresponding aspect of each leaflet (Davachi F, et al, 1971).

The majority of chordae which attach to the leaflets form a fine network of delicate threads that spring from larger chords near the papillary muscles, but some chords run singly and non-branching from the papillary muscles to the leaflet (Perloff JK, and Roberts WC, 1972).

Chordae tendineae to each cusp are arranged in a constant order into three sets :-

1. Two major fixing chordae arising from separate heads of the same papillary muscle or from two different muscles, in case of the anterior and posterior cusps, each major fixing chordae gives three sets of branches:-
 - a- The proximal branches, attached to the ventricular surface of the leaflet near the annulus.
 - b- The intermediate branches, attached to the ventricular surface of the leaflet near the edge.
 - c- The marginal branches, attached to the edge of the leaflet.
2. Two lateral fixing chordae, give several branches attach to the lateral border of the cusps at the edge or just near it.
3. Two commissural chordae, attached to the adjoining sides of two leaflets, and have a characteristic appearance because of the mode of their branching which is regular with alternate branches attaching to one of the adjoining cusps

(Kalmanson D, 1976)

The rough zones of the anterior and posterior leaflets and the basal zone of the posterior leaflet receive the insertion of chordae tendineae (Lam JHC, et al, 1970).

Each rough zone chorda splits into three chords soon after its origin from the papillary muscles, one inserts into the free margin of the leaflets, one inserts beyond the free margin to the line of closure, and an intermediate cord inserts between the two. There may be further branching giving rise to secondary branches which insert in the same sites (Kalmanson D, 1976).

From the anterior leaflet rough zone chordae, there are the strut chordae, which are two thick and large chordae that originate from the tips of the postromedial and antrolateral papillary muscles and inserting between four and five O'clock position on the postromedial side and between seven and eight O'clock position on the antrolateral side in about 90% of the hearts (Lam JHC, et al 1970).

The rough zone chordae of the posterior leaflet are shorter, and thinner than those of the anterior leaflet, and the posterior leaflet does not have strut (thick) chordae.

The chordae to the antrolateral commissure and the adjoining half of the anterior and the posterior leaflets arise from the antrolateral papillary muscle group. Those pass to the postromedial commissure, and the adjoining half of the anterior and posterior leaflets originate from the postromedial group of papillary muscles.

There is no significant difference in the total number of chordae between males and females, there are about 25 chordae

insert into the mitral valve leaflets, nine into the anterior leaflet (seven rough zone and two strut chordae), 14 into the posterior leaflet (ten rough zone, two cleft, and two basal chordae) and two insert into the commissures (Kalmanson D, 1976).

There may be fleshy and muscular chordae occasionally arising from the antrolateral papillary muscles in the normal hearts. (Lam JHC, et al, 1970).

The attachment of the chordae to the head of each papillary muscle is constant :-

- * Major fixing chorda to the anterior cusp.
- * Lateral fixing chorda to the anterior cusp.
- * Commissural chorda to the anterior and posterior cusps.
- * Major fixing chorda to the lateral or the medial cusp.
- * Commissural chorda between the lateral or the medial cusp and the posterior cusp.
- * Lateral fixing chorda to the posterior cusp.
- * Major fixing chorda to the posterior cusp.

(Kalmanson D, 1976).

Classification of the chordae tendineae has a significant bearing on the syndrome of ruptured chordae (Ranganathan N, et al, 1970). Rupture of strut chorda will result in severe regurgitation and a flail anterior leaflet, while rupture of one or more branches of the rough zone chordae especially of the posterior leaflet result in only a moderate regurgitation

The papillary muscles

There are two groups of papillary muscles in the left ventricle, antrolateral and postromedial groups (Roberts WC, 1983) each group gives attachment to the chordae of one half of both mitral leaflets. (Kalmanson D, 1976).

The postromedial group is smaller than the antrolateral one (Roberts WC, 1983).

Morphologically there are three types of the papillary muscles;

- a. A papillary muscle fully adherent to the adjacent ventricular myocardium and protruding very little into the ventricular cavity, with free trabecular attachment.
- b. A papillary muscle with on third or more of the body protruding freely into the ventricular cavity with very few or no trabecular attachment.
- c. An intermediate type.

(Ranganathan N, and Burch GE, 1969).

The blood supply to the antrolateral papillary muscle is through branches from the anterior descending coronary artery and either the diagonal left ventricular arteries or the marginal branches of the left circumflex artery (Kalamanson D, 1976) And that of the postromedial papillary muscle is through branches from the left circumflex artery and or branches of the right coronary artery. (Ranganathan N, and Burch GE, 1969).

Two main types of arrangement of the long penetrating intra-

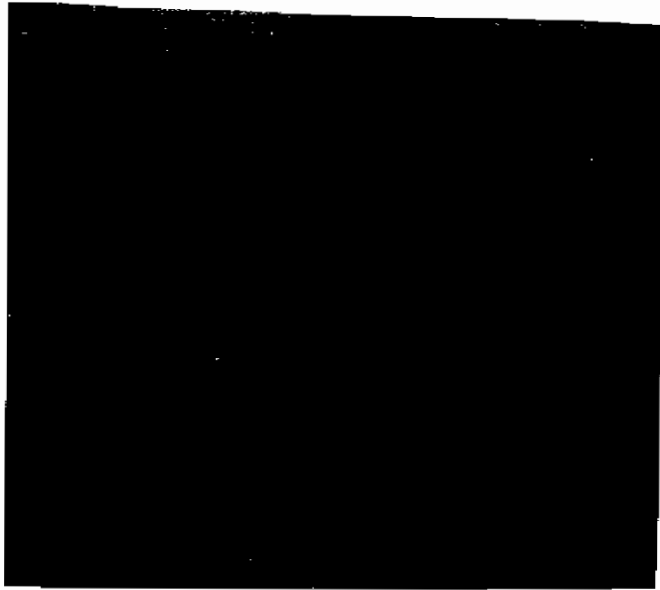


Fig. 1 Normal mitral valve.
(Ruckman RN, and Van Praagh R, 1978)

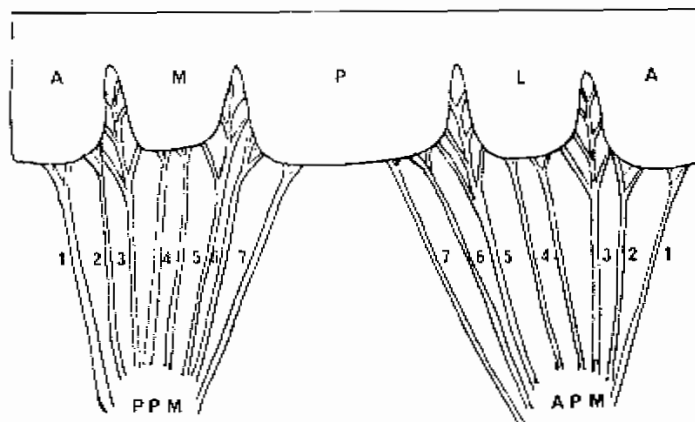


Fig. 2 Diagram of mitral valve showing the arrangement of the chordae (1) Major fixing chorda to anterior cusp (2) Lateral fixing chorda to anterior cusp (3) Commissural chorda (4) Major fixing chordae of lateral and medial cusps (5) Commissural chorda (6) Lateral fixing of posterior cusp (7) Major fixing of posterior cusp.
(Kalmanson D, 1976)

EMBRYOLOGY OF THE MITRAL VALVE

The mitral and tricuspid valves arise from an endocardial proliferation in the primitive atrioventricular canal which separates the common atrium and ventricle, the dorsal and ventral endocardial cushions fuse to divide this canal into two orifices (Cleland W, et al, 1969).

Valve formation takes place only after completion of ventricular septation. (Wenink ACG, and De Groot ACG, 1982).

Early in the developmental stage, the left atrioventricular orifice is surrounded medially by tubercles of the anterior and posterior endocardial cushions, and laterally by the left lateral cushion (Ruschhaupt DG, et al, 1976).

In the definitive heart the trabeculations in the left ventricle reach the mitral valve along the entire parietal wall of the base of the ventricle.

Trabeculation and undermining beneath the endocardium extend from the greater curvature towards the atrioventricular canal, to the base of the anterior and posterior atrioventricular cushions, and towards the atrium, cutting between the atrioventricular cushions, and the parietal wall of the base of the left ventricle.

Free flaps consisting of the atrioventricular cushion mesenchymae on their atrial side and ventricular trabeculations on their ventricular side protrude into the cavity of the left ventricle, these flaps are primordia of the mitral leaflet, and

the trabeculations are the foreunners of the chordae and papillary muscles (Goor DA, 1975).

Valve leaflets formation occurs late in development (De Groot ACG, and Wenink ACG, 1984) and the atrioventricular valve cusps are not all formed at the same time, the anterior cusp is the first to develop followed by the posterior one (Eurst JW, et al, 1982).

The anterior leaflet of the mitral valve is formed from the fusion of the anterior and the posterior atrioventricular cushions, and the trabeculating processes along the diaphragmatic and the anterior walls of the left ventricle provide this leaflet with insertions into the posterior and anterior papillary muscles.

The posterior leaflet is derived from the lateral atrioventricular cushion, and also acquires its insertion into the two papillary muscles in the same manner (Goor DA, 1975).

The valvular primordia is composed of endocardial tissue and protrudes into the ventricle as a thick distinct valvular leaflet, then the myocardial tissue begins to infiltrate and replace the endocardial elements of the valvular primordia (Layman TE, and Edwards JE, 1967).

Early most of the valvular tissue originates from the endocardial cushions, but the chordae tendineae and the papillary muscles are derived from the myocardium. (Dালiento L, et al, 1984).