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A spectrum of normality relevant to mitral valve prolapse

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SUMMARY The intricate anatomy of the mitral valve apparatus suggests that variations in its architecture may have functional significance. In this study, therefore, deviations from a basic scheme of normality of chordal support have been documented, with particular reference to the occurrence of deficient chordae. Chordae were considered as deficient when they had an irregular branching pattern, not compensated by neighbouring chordae, leaving parts of the valve leaflets less well supported than others. One hundred ‘normal’ heart specimens were studied, in which the mitral valve in the unopened state showed a regular appearance, and compared with 40 hearts in which the valve showed a deformity, classified as ballooning in 38 cases and as prolapse in two.

Deficient chordae were identified in eight of the 100 ‘normal’ hearts and in 36 of the 40 hearts with a valve deformity. In the latter group the chordal deficiency was directly related to the leaflet deformity present. Deficiencies in chordal branching and distribution affected commissural chordae and rough zone chordae, showing a particular preference for the posteromedial commissural area and the middle scallop of the posterior leaflet. The latter abnormalities showed a tendency to be associated with finger-like posteromedial papillary muscle groups and a haphazard arrangement of chordal take-off. Both specimens with necropsy evidence of prolapse had such an arrangement.

The findings showed that there is a ‘spectrum of normality’ with respect to the anatomy of the mitral valve chordal apparatus. It provides an anatomical basis for echocardiographic recordings of disharmonious mitral valve movements, frequently recorded among otherwise healthy individuals. Moreover, it is assumed that such minor variations may play a role in the pathogenesis of the syndrome of mitral valve prolapse, by rendering unsupported parts of leaflets particularly vulnerable to high pressures.

The functional anatomy of the mitral valve apparatus has been well documented in recent years and it is at present widely acknowledged that disturbances in co-ordinated interaction of the various anatomical components may underlie valve insufficiency (Perloff and Roberts, 1972). For obvious reasons most interest in this field has been generated by the effects of myocardial ischaemia. However, considering the intricate architecture and the delicate interplay necessary for proper function, it can be anticipated that variations in anatomy may also become of importance. It is surprising, therefore, that little attention has been given to this particular aspect of the functional anatomy of the mitral valve apparatus.

When establishing a concept of ‘normality’ for the mitral valve the studies of the Toronto group of investigators may serve as a point of departure (Lam et al., 1970; Ranganathan et al., 1970). They presented a scheme and classification for the normal arrangement of both mitral valve chordae and leaflets and mention that variations in morphology were not infrequent. Our initial experience was in keeping with their basic scheme but we became particularly aware of the potential functional significance of ‘minor’ variations in chordal distribution encountered within their basic scheme of normality. The nature of some of these variations was such that some parts of the mitral valve leaflets seemed less well supported than others. It seemed that this observation could be of significance regarding mechanisms for mitral valve prolapse, since echocardiographic studies indicate that abnormal valve movement is often found among apparently healthy subjects (Markiewicz et al., 1976; Procacci et al., 1976). In view of these
considerations, the mitral valve apparatus has been studied in 140 necropsy specimens, in an attempt to evaluate the significance of variations in chordal distribution for the occurrence of deformities in the valve leaflets.

**Subjects and methods**

Hearts were obtained from necropsies on patients who had died from causes unrelated to cardiac disease. In none had a mitral valve disorder been noticed during clinical examination, but they had not been seen by a cardiologist.

Hearts were excluded which showed necropsy evidence of rheumatic or infectious valve diseases, calcified mitral ring, or conditions known to affect intrinsically the connective tissues, such as Marfan's disease or mucopolysaccharidoses.

The study was based on 140 heart specimens, examined in the fresh state. In all instances the left ventricle was filled with tap water, through a cannula in the aortic ostium, after tying the ascending aorta. The pressures mounted to an approximate 200 mmHg. The hearts were then divided into two discrete groups. The first group consisted of 100 mitral valves which, when observed from the left atrium with the left ventricle unopened, neatly closed the mitral orifice. Usually, such leaflets show hoods, defined as an upward bulging of interchordal leaflet tissues. The degree of hooding may vary from one individual to the other, but there is a tendency for them to be more pronounced in the elderly and in conditions that lead to left ventricular hypertrophy. This suggests that hooding is an acquired phenomenon rather than a basic aspect of 'normality'. However, because of the occurrence of hoods in nearly all hearts this finding was considered 'normal'. The 'normal' hearts were obtained from 62 male and 38 female patients; the ages varied from 23 to 92 years, with an average of 61 years.

The second group consisted of 40 heart specimens in which a deformity of the mitral valve was present as observed from the left atrium. The abnormality most frequently encountered was termed 'ballooning deformity', defined as the condition in which a leaflet, or part of a leaflet, showed an upward bulging extending beyond that of the 'normal' interchordal hoods. Moreover, the degree of bulging towards the left atrium was greater than that seen with hoods. However, when the two mitral valve leaflets were brought into apposition there was no actual overshoot in the postmortem state. This was in contrast to the condition in which the aforementioned procedure resulted in an actual overshoot of the free margin of the affected leaflet, which has been considered as valve prolapse. The ballooning deformities were encountered in all 40 specimens, while an additional prolapse was present in two hearts. These abnormal hearts were obtained from 23 male and 17 female patients; the ages varied from 38 to 86, with an average age of 63 years.

In each of the 140 specimens a close study was made of the architecture of the chordae, taking the basic scheme of normality defined by the Toronto group (Lam et al., 1970; Ranganathan et al., 1970) as the point of departure (see below). The study has focused in particular on the occurrence of chordae exhibiting a deficient branching pattern, without sufficient overlap of neighbouring chordae, an arrangement leaving part of a leaflet apparently less well supported than would be expected from 'normality'. The term deficient chorda will be used for this particular arrangement.

**Basic scheme of normality**

A basic scheme of 'normality' of the mitral valve was based on the work of the Toronto group of investigators (Lam et al., 1970; Ranganathan et al., 1970). They introduced a classification of chordae, relevant to mitral valve function, by distinguishing two major sets of chordae. Firstly, they introduced the term 'commissural chordae' for the cords which support the anterolateral and the posteromedial commissural areas. In the 'typical' situation there is one cord for each commissure (Fig. 1). It arises as a single main stem from the underlying papillary muscle group, and then branches into fan-like structures which insert into the free margin of the two commissural leaflets (Fig. 1C). The lateral spread of these branches is wider in the posteromedial commissural area than it is at the anterolateral commissure. The commissural chordae are considered to play a major role in the process of folding and unfolding of the leaflets bordering on the commissure. Lam and associates (1970) mentioned the absence of one anterolateral commissural chorda among their 50 hearts, but they did not expand on the occurrence of 'atypical' commissural chordae with a deficient branching pattern.

The second set of chordae, as defined by the Toronto workers, is formed by a group of 'leaflet chordae'. Within this group the 'rough zone chordae' are the ones which give the major support to both the anterior and posterior leaflets. In the 'typical' situation each 'rough zone chorda' divides into three separate cords, shortly after the origin of the main stem from the papillary muscle (Fig. 1E). One of these cords inserts into the free margin of the leaflet, while the second and third cord insert beyond the free margin, reinforcing the site of the
line of closure. Among the rough zone chordae for the anterior leaflet there is a thickened 'strut' chorda, one from each papillary muscle inserting into the corresponding half of the leaflet. The rough zone chordae are considered to be of major significance in maintaining the integrity of the valve during systole and should therefore exhibit a regular distribution over the anterior and posterior leaflets. However, Lam and associates (1970) reported the occurrence of so-called 'atypical' rough zone chordae, which they defined as cords exhibiting a deficient branching, albeit that at the sites of insertion an overlap from neighbouring chordae took place in a high percentage of cases. These investigators observed 'atypical' chordae among 37 of their 50 hearts.

The cleft chordae, delineating the clefts which accentuate the scallops of the posterior leaflet, were considered as a separate entity within the group of leaflet chordae. They insert into the free margin as
well as the rough zone areas of the two scallops bordering on the cleft, so that from a functional point of view they probably act like rough zone chordae (Fig. 1D). For the purposes of this study we have included them in the latter category. Basal chordae constitute the final type within the group of leaflet chordae, present only for the posterior leaflet. These cords originate from the ventricular free wall and usually fan out just before their insertion into the basal aspect of the posterior leaflet, close to the annulus fibrosus. This arrangement suggests that they reinforce the basal part of the leaflet during systole, but their immediate functional significance seems limited. Lam and associates (1970) found basal chordae in only 31 of their 50 hearts, while their number and location showed considerable variations. For these reasons we have not included basal chordae in our studies.

Results

Of the 100 normal hearts there were 61 in which the basic arrangement of the chordae was designated as normal; 39 hearts showed a variation within the basic scheme. In 31 of these 39 specimens a sufficient overlap of neighbouring chordae was present, leaving a total of eight normal hearts with deficient chordae. In all of these eight hearts the deficient chordae supported the anterior leaflets.

Of the 40 hearts with a pronounced valve deformity, all showed a variation in chordal arrangement, which in four instances seemed to be compensated by neighbouring cords. In other words, 36 of these 40 hearts (90%) showed deficient chordae. In all instances the observed deficiency showed a direct topographic relation with the valve deformity. In the four hearts where no deficient chordae could be identified, we observed ballooning deformity of the middle scallop of the posterior leaflet. The deficient chordae were distributed as follows.

Commissural chordae

Deficient commissural chordae were identified only among the 40 hearts with a grossly identified leaflet abnormality. In each instance the valve had a ballooning deformity. Nine specimens presented such a deficient chorda, affecting the posteromedial commissure in seven and the anterolateral commissure in two cases.

Two major aberrations in architecture were identified. The first abnormality was characterised by a commissural chorda which, after a basically normal origin, showed an irregular and deficient branching pattern. This particular arrangement in itself was not infrequent among normal hearts either, but sufficient overlap from rough zone chordae restored a regularly distributed support. The commissural chordae reported here lacked such additional support, rendering part of the leaflets relatively unsupported (Fig. 2A and B). The second form was characterised by a profoundly shortened chorda, showing only sparse ramifications which inserted at the site of the deepest indentations of the commissure. Thus an extensive area of free margin was left without chordal insertions (Fig. 2C and D).

Superficial examination of these chordal aberrations may suggest a similarity with a post-rheumatic process (Fig. 2A and D). However, close inspection will reveal that most chordae are slender or fan-like, and that the main abnormality is not one of chordal fusion but of chordal deficiency. The associated valve leaflet shows a localised tissue response composed of scar tissue, but such reparative processes are in themselves non-specific and cannot be interpreted as favouring a rheumatic origin.

Rough zone chordae

Deficient rough zone chordae were present in eight of the 100 normal hearts and in all 40 hearts showing a valve deformity. Such anomalies affected both the anterior and posterior leaflets.

Anterior leaflet

In all the eight ‘normal’ valves in which deficient chordae were detected, these had supported the anterior leaflet. The lateral and medial halves of the anterior leaflet were involved in three and five instances, respectively. Of the 40 deformed valves, 26 showed such a deficiency in chordal distribution for the anterior leaflet. In nine instances the lateral half of the leaflet was involved, while in 17 cases the anomaly related to the medial half.

Between both groups there were no major differences regarding the type of chordal anomaly. Basically, the deficiency encountered was caused by one of two major aberrations. Firstly, a strut chorda was present, but it was not accompanied by the usual neighbouring slender rough zone chordae, leaving a ‘bare’ area not supported in the usual way (Fig. 3A and B). In some instances this thick and deficient strut chorda contained muscle extending from the papillary muscle. The chorda could thus be designated as a persistent muscular chorda, but its categorisation in this series was based on its deficient branching pattern rather than its specific build. The second type consisted of an irregular grouping of slender chordae, usually crowding close to the free margin of the leaflet, leaving the more distal part of the ‘rough zone area’ with less chordal

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support than expected from the basic scheme of normality (Fig. 3C and D).

Posterior leaflet
Deficient chordal support for the posterior leaflet was seen only among the 40 cases with a mitral valve deformation. It should be noted that the number of scallops varied considerably, since only 30 of the 40 specimens showed a tri-scalloped posterior leaflet. Of the remaining 10 specimens, eight showed four scallops while two specimens had a total of five scallops. However, a 'middle' scallop could always be identified because of a mutual chordal support derived from both papillary muscle groups. For clarity all leaflet tissues bordering the middle scallop will be grouped together as either the 'postero-medial commissural scallop' or the 'anterolateral commissural scallop', according to the terminology of Ranganathan et al. (1970). Abnormalities of cleft chordae will not be classified separately.

Deficient chordae were present in 36 instances, affecting only the middle scallop in 15 instances, only the posteromedial commissural scallop in two cases, and affecting both scallops at the same time in 19 hearts. The anterolateral commissural scallop was not involved in any of these specimens. In all instances ballooning was present, while in two hearts additional prolapse was identified. The latter deformity affected the middle scallop in both cases.

Three major forms of chordal aberration were identified. The first variety was mainly characterised by an overall irregular arrangement of chordae, which affected both their mode of origin as well as their insertion. This irregular arrangement was frequently associated with an underlying postero-medial papillary muscle group of a 'scattered' type with multiple small heads with muscular or fibrous bridges between them. Most chordae originated in a rather haphazard fashion from this intricate composition, while others originated from the posteoinferior free wall of the left ventricle (Fig. 4). Twenty-five of the 36 specimens with deficient chordae showed this architecture.
Fig. 3 Examples of deficient rough zone chordae for the anterior leaflet. (A) A thickened strut chorda (asterisk) for the lateral half of the anterior leaflet, while the remaining rough zone chordae insert into the free margin, leaving the area neighbouring the insertion of the strut chorda relatively unsupported. The part of the leaflet so affected shows mild ballooning (arrows). Note the irregular chordal branching pattern for the medial half of this leaflet.

(B) The undersurface of the anterior leaflet in another specimen. Again a thickened strut chorda (asterisk) is present, but the immediate surroundings are devoid of chordal insertions. There is an upward bulge of the leaflet in this region.

(C) A bundle of rough zone chordae derived from the posteromedial papillary muscle group (PM) which insert into the free margin of the medial half of the anterior leaflet, leaving the more basal aspect unsupported. A ballooning deformity is present at that site (arrows). An irregular chordal distribution pattern is present also for the lateral half of this leaflet.

(D) A similar situation, in greater detail, for the lateral half of the anterior leaflet. Note that both chordae and leaflet at that site are thickened.

A second form of deficient chordae was characterised by pillar type papillary muscle groups, with most chordae originating from the tips. Those for the support of the ‘middle scallops’ were long and slender and inserted close to the free margin of the leaflets, lacking the typical distribution pattern of rough zone chordae. This arrangement was present in seven hearts, and included one of the cases having necropsy evidence of prolapse (Fig. 4B).

A third major form was characterised by the presence of a single, ‘isolated’ long chorda, originating as a separate structure from the posteroinferior free wall of the left ventricle. This chorda then inserted into the overlying scallop, not accompanied by other chordae for additional support. This peculiar anatomy was encountered in four specimens and in each of these the anomaly involved the middle scallop. One of the two specimens having valve prolapse presented this type of chordal arrangement (Fig. 4C and D).

Discussion

The intricate architecture of the mitral valve apparatus, in which a delicate and co-ordinated interaction of various anatomical components is a necessity for proper function, is well recognised (Perloff and Roberts, 1972). As a consequence,
there is an increasing awareness that a multitude of different disease processes, by interfering with such a co-ordinate action, may act through a final common pathway and result in mitral regurgitation. Papillary muscle dysfunction can be regarded as one such example (Cheng, 1969; Shelburne et al., 1969; Perloff and Roberts, 1972). A similar awareness is growing regarding the syndrome of mitral valve prolapse. The floppy valve, defined as expansion of the mitral cusp area with elongation of chordae, is considered a major cause of this syndrome, with weakness of the central cord of the valve leaflet as the essential lesion (Davies et al., 1978). Since valve leaflets so affected almost always show mucoid changes in the central connective tissue core, it has been proposed that this abnormality constitutes the essential lesion (Jeresaty, 1973, 1975; Davies et al., 1978). However, mitral valve prolapse has been documented under quite different clinical circumstances, though most of these in some way or other interfere with a co-ordinated interaction of the anatomical ‘building blocks’ of the mitral valve apparatus (Barlow et al., 1968; Perloff and Roberts, 1972; Bulkley and Roberts, 1975). There is a tendency, therefore, no longer to regard mitral valve prolapse as an entity in itself, but rather as an expression of a number of potential causes (Nutter et al., 1975; Aranda et al., 1976; Devereux et al., 1976; Lesch, 1976). Among those causes a disproportion in size between the mitral valve and the left ventricular cavity has been suggested as a potential mechanism (Jeresaty, 1971; Criley and...
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Kissel, 1975; Cheng, 1976). Perloff and Roberts (1972), in their excellent display of the functional anatomy of the mitral valve apparatus, also mention the possibility that ectopically inserted chordae may contribute to mitral regurgitation, referring to a report by Levy and Edwards (1962) on the occurrence of ectopic ventricular chordal attachments as a cause of impaired valve function. These suppositions are of considerable interest because they introduce a concept of individual anatomical variations, themselves not necessarily abnormal, which underlie a functional disorder. Such a concept is appealing, not least because echocardiographic studies have disclosed a considerable variation in the occurrence and duration of mitral valve prolapse (Higgins et al., 1976) and a surprisingly high prevalence among apparently healthy individuals (Markiewicz et al., 1976; Proacci et al., 1976). One may indeed wonder whether these sophisticated techniques, rather than showing overt disease, do not also record individual variations in anatomy, a proposition also favoured by Markiewicz and associates (1976) when discussing their own figures.

It is of interest, therefore, that so little attention has been given to the variability of the mitral valve apparatus. In a recent work on the normal anatomy of the mitral valve, concerning the build of leaflets and chordae tendineae, it is mentioned that aberrations from a 'basic scheme of normality' do occur (Lam et al., 1970; Ranganathan et al., 1970). In fact, these investigators mentioned that 'atypical' chordae were present in 39 of the 50 hearts studied. However, in approximately 60 per cent of these instances the deficient branching pattern of these chordae was compensated by an overlap from neighbouring chordae, a percentage that none the less left approximately 12 out of their 50 hearts with some sort of deficient local support for part of the valve leaflets. If one considers the fact that the mitral valve sustains considerable pressures for a prolonged time, one may perhaps speculate on the effects of an irregular distribution of chordal support on leaflets. In particular, it is already known that atrioventricular valves have a profound tendency to develop distinct upward bulges of the interchordal leaflet tissues with increasing age of the patient and in conditions accompanied by a prolonged rise in intraventricular pressures. This phenomenon has been termed 'hooding'. Indeed, Oka and Angrist (1961) have suggested that valve deformities, similar to the ones we have defined as 'ballooning deformities', result from ageing. Study of the core of the leaflets under those conditions will reveal an increase in mucopolysaccharides, probably an expression of tissue injury. Moreover, Pomerance (1969) has shown that the incidence of such deformities in a random necropsy series was encountered much more frequently in patients over 50 years of age than in younger individuals. This is not surprising if one is willing to accept that 'deficient chordae' may be present among valves which otherwise 'look normal'. If the anatomy is such that the interchordal areas widen, for instance because of a deficiency in the pattern of chordal branching, valve deformities can be expected with time. One could then also speculate whether this train of events might lead to a valve cusp with a weakened central core, expansion of the cusp area, and elongation of chordae: in other words, are some 'floppy valves' actually the end result of primary deficient chordal support?

This study has shown that pronounced differences occurred between a group of hearts designated as 'normal' and a group having a valve deformity. Among 100 'normal' hearts we found 39 specimens with a variation within the basic scheme of normality proposed by Lam et al. (1970) and Ranganathan et al. (1970). We also found that most of the variations in the present series had no deleterious effect on leaflet support because of overlap from neighbouring chordae. In this series of 'normal' hearts, only eight showed a 'deficient chorda' defined as a chorda with a non-compensated atypical branching pattern. However, it should be pointed out that this series of 'normal' hearts was selected on the basis of not exhibiting a valve leaflet deformity when observed in the closed state from the left atrial cavity. Hearts with such a deformity were included in a separate group and the incidence of deficient chordae was compared with that in the group of normal hearts. The results disclose that deficient chordae were detected among 36 of the 40 mitral valves showing a leaflet deformity. In all instances the area of the leaflet showing the deformity corresponded with the site of the deficient chorda. These findings, therefore, suggest that 'minor' variations in architecture of the chordal apparatus may leave some parts of the leaflets less well supported than others, a phenomenon which could then result in a deformity of the leaflet at that particular site. These abnormalities showed a definite tendency to occur at the site of the postero-medial commissure and related parts of the posterior leaflet, including the middle scallop, in contrast to the anterolateral parts of the valve apparatus which were less frequently affected. This observation is particularly valid when one is dealing with a 'forme frust' of a developmental anomaly, since the postero-medial papillary muscle complex was often intricately related to the chordal anomalies observed. The middle scallop in some of these
hearts was reminiscent of a sort of ‘watershed area’, showing chordae from both papillary muscle groups inserting in the lateral free margin of the scallop, leaving the middle part less well supported. In other specimens the middle part was sustained only by a single ‘strut-like’ chorda, leaving a large area of the middle scallop without sufficient support; in both specimens with prolapse the middle scallop was involved. One case had a preponderance of chordal insertions among the lateral free margins, while the second case showed the ‘isolated’ chordal variety. It should be re-emphasised that in both instances valve prolapse was defined from the necropsy state. Regrettably, there were no relevant clinical data on the functional state of the mitral valve, since one patient died in a traffic accident, while the other patient was seen clinically at the end-stage of a malignancy.

The present observations lead to the following assumptions. Firstly, they may provide an anatomical substrate for disharmonious valve motions observed with echocardiographic techniques, suggesting that the high frequency of valve prolapse observed among apparently healthy individuals (Markiewicz et al., 1976; Procacci et al., 1976) reflect a spectrum of normality, rather than abnormalities of the valve itself. Similarly, the presence of deficiencies in chordal distribution may render a valve vulnerable not only to sustained high pressures but also to various conditions which may affect different components of the mitral valve apparatus. Weakening of the central core of the leaflet may act as a ‘final common pathway’ to a process of chronic injury, and some cases of ‘floppy valve’ may fit within this category.

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References


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