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A histological study of the atrioventricular junction in hearts with normal and prolapsed leaflets of the mitral valve

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SUMMARY The mitral annulus is the point at which the atrial and ventricular walls meet the base of the mitral valve cusps. The suggestion that a variant of this arrangement termed "disjunction" was associated with prolapse of the leaflets prompted examination of the mitral atrioventricular junctions in seven normal hearts and six with prolapse owing to floppy mitral valves. A complete cord-like ring of connective tissue that encircled the atrioventricular junction and into which the three components were inserted at the same point was found in only one heart. The remaining hearts all showed a mixture of segments in which either the three components were inserted into a cord or simply met. Disjunction, defined as a separation of the atrial wall-mitral valve junction from the other component, the left ventricular wall, can occur both with and without a cord-like annulus. There was no significant difference in the number of segments around the left atrioventricular junction which showed disjunction in hearts with normal or prolapsing leaflets.

The feature termed disjunction is an anatomical variation of the normal morphological characteristics of the left atrioventricular junction.

The pathogenesis of the floppy mitral valve remains controversial. Many theories have been advanced since Reid proposed a valvar origin for the mid-systolic click and late systolic murmurs.[1] Jeresaty argued[2] that there were two theories to account for floppiness of the mitral valve. The first emphasises the abnormality of the valve, whereas the second postulates myocardial involvement. Most morphological evidence accords with the suggestion that an abnormality of the valve is responsible for prolapse. Abnormality of the valve has been ascribed to a primary deficiency of the dense collagenous and fibrous layer of the leaflets[3–6] or an anatomical abnormality of the cords.[7 8]

A third possibility was proposed by Hutchins and his colleagues who suggested that a distinctive and basic anomaly of a floppy valve is disjunction of the fibrous annulus—that is a separation between the atrial wall-mitral valve junction and the left ventricular attachment.[9] They argued that this anomalous feature, through a process of hypermobility of the tension apparatus, could lead to floppiness of the leaflets. This group, however, studied only limited histological sections from a large number of hearts to demonstrate the significant association between so-called "disjunction" and floppy and prolapsed leaflets.

It seemed to us that it was important to test their hypothesis by examining in detail the entire atrioventricular junction in a smaller number of hearts. This report describes our findings.

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Patients and methods

We examined 13 hearts obtained an necropsy. The mitral valve was normal in seven (from 3 men and 4 women) and floppy in six (from 4 men and 2 women). The specimens of normal hearts came from a group aged 66–80 years (mean 71·9) and those with floppy valves came from a group aged 56–89 years (mean 69·5). The valves were classified as "floppy" if they

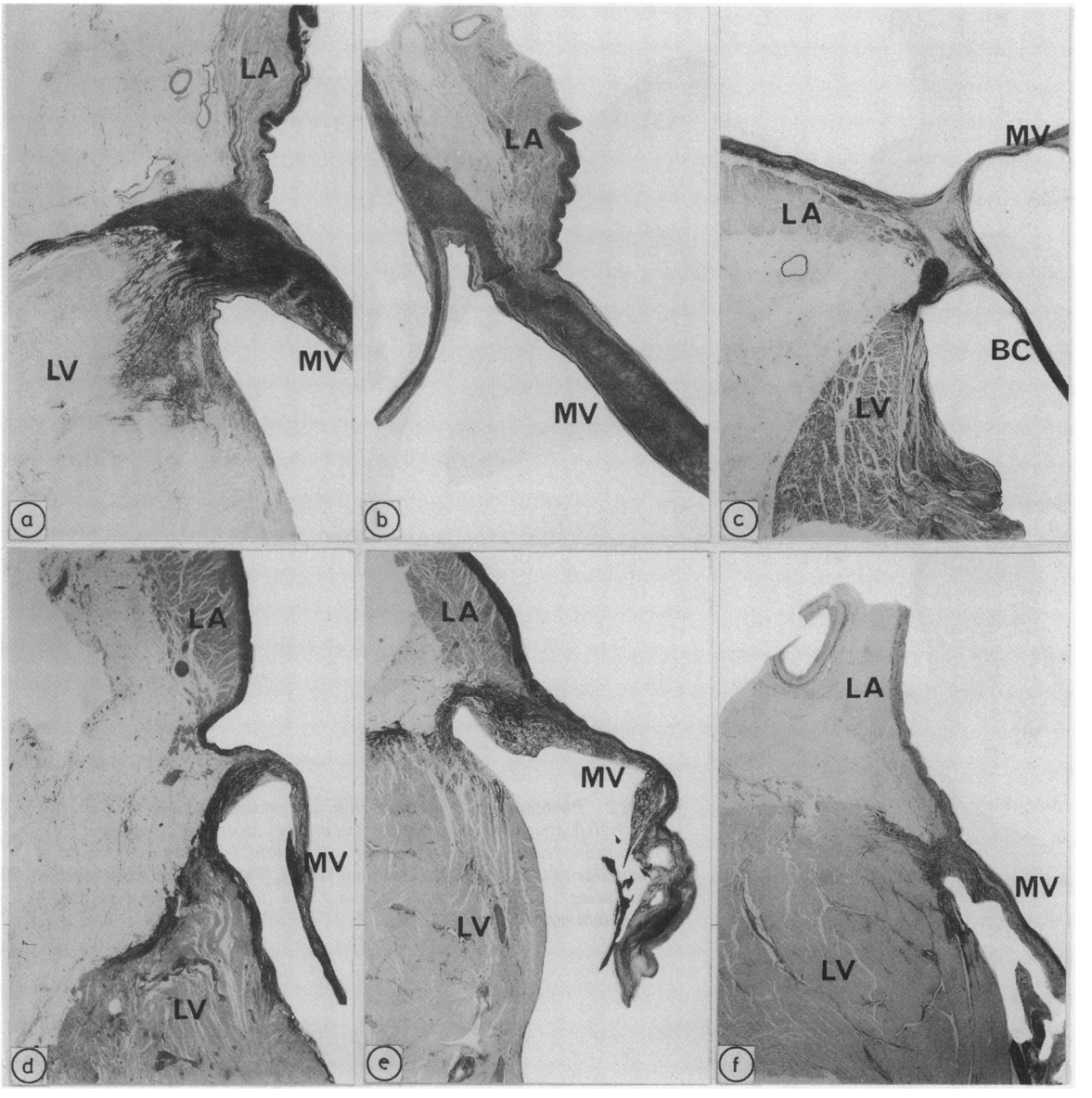

Fig 1 *Photomicrographs of sections showing structural variations in the left atrioventricular junction in hearts with normal leaflets of the mitral valve. The sections were cut perpendicularly to the atrioventricular junction. LA, left atrium; LV, left ventricle; MV, mitral valve. In (a) there is a cord-like annulus at the site of the left fibrous trigones whereas in (b) there is a fibrous continuity between the leaflets of the mitral and aortic valves. No dense fibrous aggregation resembling a cord-like annulus was detectable. In (c) a cord-like annulus supports the hinge of the leaflet from a basal tendinous cord (BC). (d) Shows a curtain-like annulus. The atrial myocardium stops short of the hinge. There is a curtain-like annulus in (e) too, where the atrial myocardium is inserted into the hinge of the leaflet. In (f) there is a curtain-like annulus attached below the apex of the ventricular wall. The arrangements seen in c, d, and e are all examples of so-called "disjunction" as defined by Hutchins* et al.[9] *Elastic-Van Gieson stain (original magnification, × 10).*

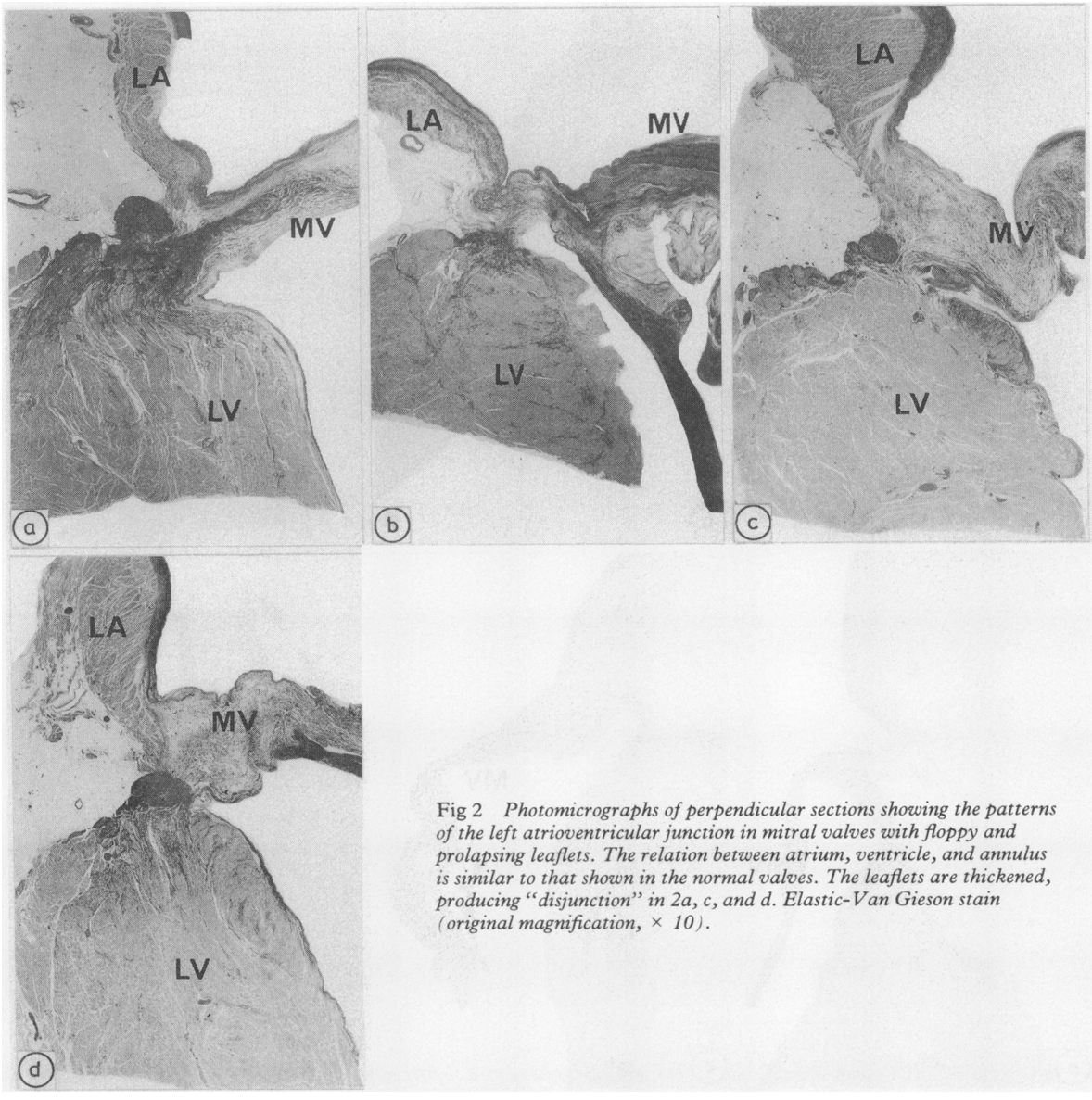

Fig 2 *Photomicrographs of perpendicular sections showing the patterns of the left atrioventricular junction in mitral valves with floppy and prolapsing leaflets. The relation between atrium, ventricle, and annulus is similar to that shown in the normal valves. The leaflets are thickened, producing "disjunction" in 2a, c, and d. Elastic-Van Gieson stain (original magnification, × 10).*

they were seen to have redundant leaflets with a dome-like expansion bulging into the left atrial chamber. The severity of the involvement of the leaflets varied. All the leaflets in all but one of the floppy valves were abnormal. In this specimen only the middle scallop of the mural leaflet was affected.

Serial sections were cut perpendicularly to the valve ring along the atrioventricular junction of each heart every 3 mm and stained with elastic-Van Gieson reagent. For each valve, we mapped the structure of the annulus and the presence of so-called "disjunction". Hutchins and colleagues defined disjunction as "a separation between the atrial wall-mitral valve junction and the left ventricular attachment".[9] We also described the fibrous support at the atrioventricular junction as cord-like or curtain-like according to its sectional structure. We have described the general region of attachment of the mural leaflet at the atrioventricular junction as the hinge regardless of whether a cord-like or a curtain-like fibrous aggregation was present. The area of fibrous continuity between aortic and mitral valves was excluded from analysis, since there is no hinge-like arrangement with the ventricular myocardium along

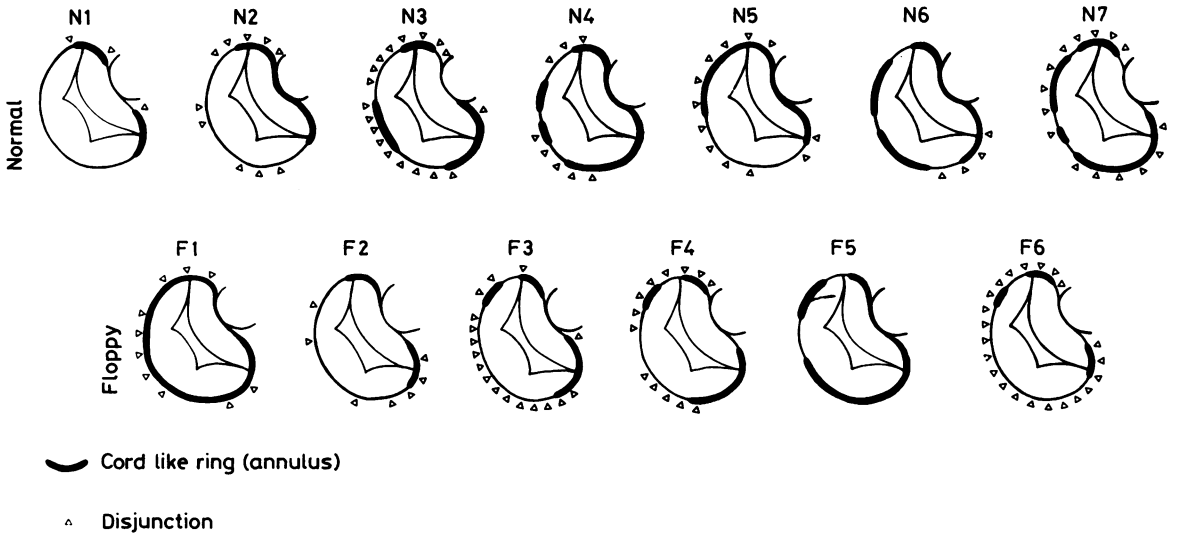

Fig 3 *Diagram showing the structural variations in the annulus around the left atrioventricular junction in all the valves studied. There is no significant difference between the number of segments showing so-called "disjunction" in the groups of hearts with normal and prolapsing leaflets.*

this segment of the junction.

Results

Examination of the histological sections showed considerable variation in the arrangement of the atrioventricular junction and in the relation of the structures making this junction, not only from heart to heart but within the same heart. Fig 1 shows the most common features.

Thick and well organised fibrous structures that produce a cord-like segment of ring are always present at the site of the left and right fibrous trigones. These are the thickened areas at the extremities of the region of fibrous continuity between the leaflets of the mitral and aortic valves. At the trigones, the aortic leaflet of the mitral valve hinges on the annulus with insertion of the atrial myocardium at the level of the hinge. The annulus itself is located at the apex of the ventricular myocardium (fig 1a). The area of fibrous continuity between the trigones is an extensive fibrous sheet joining the attachments of the two valves. Often no dense fibrous aggregation resembling a cord-like ring is detectable along this segment of valvar continuity (fig 1b). Threads of fibrous tissue occasionally extend laterally from each of the fibrous trigones and run posteriorly to embrace part of the orifice (fig 1c). In these areas, the atrial myocardium reaches the annulus, a basal tendinous cord supports the valve leaflet, and the hinge of the leaflet is displaced upwards. These extensions of cord-like fibrous tissue, none the less, do not extend as a continuous structure throughout the circumference of the valve. In most cases, the fibrous tissue of the annulus is organised in many segments of the "ring" as a thin curtain-like structure (fig 1d, 1e, and 1f). In many areas (fig 1d), the atrial myocardium stops short of the hinge of the leaflet, leaving adipose tissue to fill the gap. Alternatively, the atrial myocardium may be inserted into the hinge (fig 1e) or the annulus may be attached below the apex of the ventricular wall (fig 1f).

For the atrioventricular junction of the floppy mitral valves exactly the same range of variation is found as in normal hearts (fig 2). This holds true both for the site of abnormalities around the junction and for the type of collagenous aggregation (cord-like ring or curtain-like features). The only difference between normal and floppy valves was the presence of thickened leaflets in floppy valves. Figure 3 shows the variation in the morphology of the annulus from point to point around the junction of the hearts studied. So-called "disjunction", as defined by Hutchins and colleagues, was equally frequent in normal as in floppy valves (no significant difference as determined by Mann-Whitney test).

Discussion

Our results show that the arrangement in which a complete cord-like ring (or annulus) of connective

tissue encircles the left atrioventricular junction and supports the leaflets of the mitral valve, at the same time separating the contiguous segments of atrial and ventricular myocardium, is exceptional. Such appearances were found at limited points around the junction of all the hearts we studied. No statistical difference could be detected between normal and floppy valves. The degree of collagen in the ring, especially in its curtain-like segments, varied from an easily identifiable structure as thick as the fibrous layer of the leaflets to a thin strand of fibres.

Variation in the relation of the structures making up the atrioventricular junction was another characteristic of the mitral valve. The relation between the atrial and ventricular myocardial segments at the junction seemed closer in the floppy than in the normal valve. Indeed, some of our cases of floppy valve demonstrated the feature defined by Hutchins and his colleagues as "disjunction".[9] It seems to us, however, that there is no displacement of the junction of the myocardial segments in these hearts. Rather, the apparent separation of atrial wall, valve leaflet, and ventricular myocardium ("disjunction") is produced solely by the thickness of the leaflets at the hinge. It is, therefore, difficult for us to accept the view that in floppy valves there is a significant association with "disjunction" of the atrioventricular junction.

It should be noted that in the study of Hutchins and his colleagues only one block was taken for histological examination from the obtuse margin of the mitral annulus of each normal heart.[9] This method probably accounts for the small percentage of "disjunction" found in the normal hearts studied by Hutchins *et al.*[9] We agree with Hutchins *et al* when they state that "disjunction" is an anatomical variation of the normal morphological characteristics of the mitral annulus,[9] as it is in floppy valves too. If hypermobility of the mitral apparatus is important in producing prolapse, it is probably produced by other abnormalities over and above so-called "disjunction".

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