Persistent Common Atrioventricular Canal

Anatomy and Function in Relation to Surgical Repair

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This paper was written first in 1960. At that time cases of the complete form of persistent common atrioventricular canal or endocardial cushion defect were regarded as consisting of an ostium primum defect of the atrial septum, a cleft in the anterior cusp of the mitral valve, a cleft in the septal cusp of the tricuspid valve, and an interventricular septal defect.

Partial forms of the defect had only the first two anomalies, and transitional forms had the first three. Wakai and Edwards, however, had pointed out that the ventricular septum showed an unusual concavity below the atrioventricular valves in the partial and transitional forms. Tissue resembling fused chordae might be seen filling a gap between this concavity and the valves. Accessory chordae tendineae between mitral cusps and the ventricular septum were held to hinder mobility of the sutured cleft and lead to persistent mitral insufficiency postoperatively.

It was evident from clinical, hemodynamic, and surgical studies that not all cases had mitral insufficiency. Yet the accepted surgical treatment was complete suture of the free edges of the mitral cleft and patching of the ostium primum defect of the interatrial septum. Interventricular septal defects when present were closed either by means of a patch sutured to the septum and the under surface of the atrioventricular valves or by the suture of the valves down to the top of the concavity of the interventricular septum. The cutting of so-called abnormal chordae was being practiced by some surgeons.

The application of these technics was resulting in a certain mortality (acceptable perhaps for the time although too high by modern standards) for the partial forms and a prohibitive mortality for the admittedly more hemodynamically significant complete form. In addition to this crude assessment of the worth of the procedure it was evident to the author that not all of the cases of the partial form were relieved of their mitral insufficiency, and some without clinical evidence of mitral insufficiency preoperatively had it postoperatively. Clinical and hemodynamic confirmation of this opinion appeared around this time, and more evidence has since accumulated.

Material and Methods

An anatomic and functional study of the mitral valve was made in the course of developing a mitral valve prosthesis and further studies have since been made. The anatomy and function of the mitral valve were analyzed in cases of persistent common atrioventricular canal as seen in a collection of 12 postmortem specimens at the Mayo Clinic and in seven cases operated on by Dr. John Kirklin during a 3-month period, and further observations on six postmortem hearts and 14 cases operated on have been made in Cape Town.

Normal Mitral Valve

The features of normal mitral valve anatomy and function relevant to this discussion are as follows.

Anatomy

The Interventricular Septum

The posterior margin of this septum is related superiorly to the root of the aorta. More specifically, this relationship is to half of the base of the noncoronary sinus of Valsalva and...
the whole of the base of the right coronary sinus of Valsalva. The inferior half of the margin of the septum is joined to the interatrial septum; this junction taking place along a straight line between the meeting point of the septa and the left and right atrial and ventricular walls inferiorly and the meeting point of the noncoronary sinus of Valsalva and the two septa superiorly (fig. 1A).

**Figure 1**

A, top. Normal heart. Left ventricle opened through posterior cusp. Most of left ventricular wall retracted upwards and forwards. The noncoronary sinus of Valsalva has been exposed. The white pinheads mark the borders of the interventricular septum. The upper arrow points to the junction of the noncoronary sinus of Valsalva and the interventricular and interatrial septa. The lower arrow points to the junction of the septa and the ventricular and atrial walls. B, bottom. Partial form of common atrioventricular canal. Dissected as above. This patient died during digital exploration in 1952. Cardiac catheterization had revealed no mitral insufficiency. NC, noncoronary sinus of Valsalva; A, anterior mitral cusp; C, commissural tissue.

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**The Mitral Cusps**

There are two main cusps separated and joined by two areas of commissural tissue. The anterior cusp reaches the interventricular septum at its posteromedial border, the posteromedial commissural tissue is attached to the top of the septum and the posterior cusp is attached to the ventricular wall (fig. 2A).

**The Anterior Cusp of the Mitral Valve**

The base of the anterior cusp is attached primarily to the root of the aorta, where it is related to the whole or most of the left coronary aortic cusp and half of the noncoronary cusp. It stretches across the left ventricular outflow tract between the ventricular septum and wall (figs. 2A and 3A).

**The Chordae Tendineae of the Anterior Cusp**

The free edge of the anterior cusp has a central bare area devoid of direct chordal support. All the rest of the edge is supported by first-order chordae tendineae while the body of the cusp is supported by several second-order chordae tendineae. The chordae to the anterolateral side of the cusp arise from the anterolateral papillary muscle and those to the posteromedial edge from the posteromedial papillary muscle (fig. 3A).

**The Papillary Muscles**

The papillary muscles arise from the wall of the ventricle. One, the posteromedial, is close to the septum while the other is more lateral and anterior. Each gives origin to chordae tendineae which are inserted into the corresponding halves of the anterior and posterior cusps and the whole of the corresponding commissural tissue.

**Septal Cusp of the Tricuspid Valve**

This cusp is, like the posteromedial commissural tissue, attached to the interventricular septum but at a deeper level in the ventricle so that a part of the muscular interventricular septum actually presents to the right atrium (fig. 2A).

The junction of the interventricular and interatrial septa lies somewhere between the vertical and the horizontal. The designation,
in a general discussion, of one end of this line as anterior or superior is arbitrary.

**Function**

1. Cutting individual first-order chordae tendineae results in localized mitral insufficiency.6

2. A cut through cusp tissue out to the atroventricular ring, being unsupported along its free edges, leaks. The integrity of the cusp tissue around the ring must not be breached if continence is to be kept.

3. In a competent valve the leading edges of the opposing cusps hang to the same level in the ventricle during diastole and this relationship is maintained in systole because of the precise control of the cusps by chordae tendineae of correct length. During systole the free edges of the cusps do not rise above the plane of the atroventricular ring. When in apposition the leaflets meet over an area of tissue. Their pliability is such that no gaps are left in the line of closure and the remain-

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**Figure 2**

A, left. Normal heart. Atria opened. Interatrial septum removed. Noncoronary sinus of Valsalva exposed. The edge of the interventricular septum is marked by arrows. B, right. Partial form of common atroventricular canal (same case as fig. 1B). The cleft runs up to the septum posterior to the noncoronary sinus of Valsalva. The valve has a tricuspid appearance. NC, noncoronary sinus of Valsalva; A, anterior mitral cusp; C, commissural tissue; S, septal tricuspid cusp.

**Figure 3**

COMMON ATRIOVENTRICULAR CANAL

ing parts balloon into the atrium (fig. 4).
4. The sum of the lengths of the cusps meeting across an orifice exceeds the diameter of the orifice (fig. 4).

The Valve in Cases of Common Atrioventricular Canal

The study mentioned above revealed the following features that did not seem to have had adequate emphasis in the past.

Anatomy

The Interventricular Septum

It is a striking feature of both partial and complete forms of common atrioventricular canal that the posterior margin of the interventricular septum is scooped out (fig. 1B). Instead of a straight line from the noncoronary sinus of Valsalva to the meeting of atrial and ventricular walls, the line of this margin of the septum passes first forward from a point between the right coronary and the noncoronary sinuses of Valsalva and after reaching a variable depth sweeps backward again to the junction of atrial and ventricular walls. By contrast, that margin of the defect formed by the interatrial septum does indeed lie along the straight line from the noncoronary sinus of Valsalva to the meeting of atrial and ventricular walls. There is in fact no deficiency of septal tissue proximal to this line apart from those occasional cases with complete absence of the septum or those with separate ostium secundum defects.

The Site of the Cleft

The cleft may, of course, not divide the mitral cusp completely but in the majority of cases it does, and then always either reaches or crosses the septum.

The Mitral Valve, Figure 2B

Anterosuperior to the cleft is a cusp that stretches across the left ventricular outflow tract from the ventricular wall to the septum in rather the manner of the normal anterior cusp except that the angle it makes with the septum is nearer 90°. On the other side of the cleft, related to the septum in the same way as the normal posteromedial commissural tissue is another cusp-like structure. The cusps on each side of the cleft may be in contact or there may be a gap between them. On the ventricular wall the posterior or mural cusp is in a more or less normal position. While the anterolateral commissural tissue is well defined, the posteromedial commissural tissue is much harder to identify. There is a very short length of tissue in the angle between septum and wall between the mural cusp and the cusp attached to the septum, that could be this part of the valve. Sometimes, however, the cusp attached to the septum resembles the posteromedial commissural tissue.

The Cusp on the Aortic Side of the Cleft, Figure 3B

This structure may resemble the anterior cusp of the mitral valve in several ways. Its base is formed by the root of the aorta: however, instead of being suspended from only half of the noncoronary aortic sinus of Valsalva and most of the left coronary aortic sinus of Valsalva it hangs in addition from the other half of the noncoronary sinus and sometimes a part of the right coronary sinus too. This results in a greater part of the left ventricular outflow tract being formed by the mitral valve (with concomitant narrowing of the outflow tract) and a different disposition of this part of the valve to the mitral orifice. Medially the base of this cusp reaches the anterior end of the scoop in the ventricular

Diagram of cross-section of left ventricle, left atrium, and aorta. (a) Diastole. (b) Systole. D, diameter of atrioventricular ring; A, length of anterior mitral cusp; P, length of posterior mitral cusp; C, distance between point of chordal origin to free edge of cusp; B, distance between point of chordal origin and plane of atrioventricular ring; X-Y, level to which cusps descend in the ventricle.

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cases examined at operation in which the cleft did not reach the septum there were no chordae tendineae between septum and cusp.

The Cusp Posteroinferior to the Cleft

This structure is basically similar to its opposite number on the other side of the cleft, i.e., it is continuous with the septal cusp of the tricuspid valve; it may be directly attached to the scoop of the ventricular septum (most often), or attached by a membrane apparently composed of fused chordae tendineae or attached by chordae tendineae with definite spaces between them; and, finally, chordae arising from the left side of the ventricular septum and inserting into the medial end of the cleft margin are often found in cusps not directly attached to the septal scoop. Posteriorly, first- and second-order chordae tendineae arise from the posteromedial papillary muscle and insert into this margin of the cusp. As already noted the disposition of this cusp around the atrioventricular ring is the same as that of the normal posteromedial commissural tissue. Indeed, while at times this cusp resembles half of a normal anterior cusp at other times it looks more like slightly elongated commissural tissue.

The Posteromedial Papillary Muscle

This entity tends to be less well developed and situated deeper in the ventricle than the anterolateral one.

The Tricuspid Defect

This defect is not the main concern of this article and is therefore described in less detail. The septal cusp is almost always deformed although it may not be split. The cleft when present is at the junction of the anterior third and posterior two thirds of the cusp and an actual deficit of tissue here is common. It is often hard to decide where the division between mitral and tricuspid tissue falls in the cusp tissue posterior to the cleft. The papillary muscle of the conus is generally poorly defined or absent.

The above description was based on the Mayo Clinic material. Further experience with postmortem and clinical material in Cape
Town has confirmed its validity and the extensive and thorough embryologic and anatomic descriptions of Los and van Mierop and co-workers are substantially in agreement with it.

**Function**

With the partial and transitional forms of common atrioventricular canal the abnormal mitral valve is usually slightly incompetent or normal in function and only rarely grossly incompetent. Although the integrity of the valve tissue around the ring has undoubtedly been breached the cusp tissues bordering the cleft generally are so arranged as to function in the way the normal cusps do. There is usually enough of them that the sum of their lengths exceeds the gap that they have to close. Their attachment to the scoop of the septum tends to make them face each other. Their leading edges hang to the same level in the ventricle and their movements are controlled by normal chordae tendineae laterally and by their attachment to the deepest part of the scooped out ventricular septum medially, so that the edges cannot rise proximal to the atrioventricular ring in systole. It is evident often that these cusps are meeting over an area of their tissue rather than along the edges of the cleft (fig. 6). When the cusps are directly attached to the scoop of the septum there tends to be poor ballooning of the cusps medially so that the breach in the continuity of valve tissue is not compensated for and there is often some insufficiency at this area (fig. 5). Where septal chordae tendineae are present the cusps are at the same time freer to balloon and yet better tethered medially and there may be no insufficiency. When the cleft involves only a part of the anterior cusp without reaching the septum the tissues on each side of the cleft are more or less in the same plane, are not effectively tethered medially and rise proximal to the atrioventricular plane in systole. None of the conditions for competent function exists and in the three cases seen by the author quite marked incompetence was present. In some cases with complete clefts there is a deficiency of tissue and then there is always some incompetence. This is commonly so in the presence of a functioning ventricular septal defect deep to the atrioventricular valves (fig. 7). The reason for this is, at least in part, the presence of another anatomic departure from the normal, not so far mentioned in this article, but previously described by van

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**Figure 6**

Partial form of common atrioventricular canal. Same specimen as in figures 1B, 2B, and 3B. View from the ventricular apex with the valve in the closed position. The arrow points to the most medial part of the cleft where its edges are unsupported and there is an area of potential insufficiency.

**Figure 7**

Complete form of common atrioventricular canal. With the ventricle closed the anterior cusp halves will meet well but there is some tissue defect at the septum.
Mierop et al. This is a widening of the septum, so that the anteroposterior diameter of the heart at the level of the atrioventricular plane is greater than normal (fig. 1A and B). The cusps are just not ample enough to cover this increased distance.

Surgical Treatment

It is evident from the foregoing description that indiscriminate and complete suturing of all clefts is not sound. In the first place there can be no point in joining together two competently functioning cusps. (The orifice must be narrowed by this procedure and can actually be made stenotic.) Then, since the placing of sutures inevitably uses up tissue, the sutured cusp may be shortened across its width and lose some mobility. The fact that the two cusp halves, because of their attachment to the scoop of the ventricle, are not in the same plane increases this possibility. The suturing of a prosthetic patch to close the so-called atrial septal defect may also use up tissue, this time at the base of the cusp, which can thus be shortened longitudinally. The carelessly placed rim of a thick prosthetic patch may press on the base of the anterior cusp shortening it and interfering with its mobility. When there is a tissue deficit at the cleft, this must of course be closed but all the potential disorders of function resulting from suturing clefts are accentuated.

When there are interventricular communications deep to the cusps they are sometimes handled by suturing the cusps down to the scoop of the ventricular septum. This can only limit the length and mobility of the anterior halves.

Suturing the cleft tends to produce an anterior cusp that instead of billowing is rather flat and taut and may even have an atrial surface with a dished shape. Edwards recognized this tendency and attributed it to the presence of “abnormal” chordae tendineae that hold the sutured cusp down to the septum. However, most clefts with chordally supported edges in fact function well. It is just this type of cleft that does not need suturing and just this situation in which attempts to improve on nature may have the opposite effect. If the chordae are cut the cleft edges will certainly be more mobile: they will in fact evert. If their “kissing edges” are stitched together, as long advocated by Kirklin, then no harm may result; but if their free edges are stitched then the new anterior cusp may well overshoot the posterior cusp. Schrire et al. recorded several cases in their series in which insufficiency was noted postoperatively. Hemodynamically significant mitral insufficiency is present postoperatively in one patient, without mitral insufficiency preoperatively, in whom so-called “abnormal” chordae were cut.

From the foregoing discussion certain rules can be formulated for the management of the mitral valve lesion in these cases.

1. The assessment of the degree of mitral insufficiency is of extreme importance. Cases fall roughly into three groups: those without insufficiency, those with hemodynamically unimportant insufficiency, and those with significant insufficiency. The first two groups are common; the third is quite rare.

2. If no insufficiency is demonstrated, the cleft must be left alone.

3. With hemodynamically unimportant insufficiency the source of the jet is determined by careful palpation and observation. Commonly it comes from just next to the septum. Only this part of the cleft then needs suturing. With partial clefts that do not extend to the septum there tends to be insufficiency over the length of the cleft, which must then be sutured completely. Some cases have a small triangular gap at the meeting of the two halves of the anterior cusp and the posterior cusp. This cannot be repaired without suturing the whole cleft and should be left alone (fig. 6).

4. Care must be taken not to do anything that can interfere with valve function. The “don’ts” may be listed as follows: (a) Do not use valve tissue to close the septal defect; the sutures used for this must be placed precisely at the junction of mitral and tricuspid tissue. (b) Do not shorten valve tissue by taking excessive bites in the repair of the
Table 1

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*This case had in addition a small ventricular septal defect.

cleft. (c) Do not distort the valve by stitching the cusps down to the scoop of the ventricular septum. This point was well made by Maloney et al.13 (d) Do not allow the edge of a stiff prosthetic patch to press down on the mitral cusps thus interfering with their mobility. (e) Do not destroy valve support by cutting so-called abnormal chordae which functionally usually do an excellent job.

5. With hemodynamically significant insufficiency a formal analysis of its causes is necessary. This involves an assessment of (a) cusp lengths in relation to orifice diameter; (b) the relationship of cusp edges to each other and to the atrioventricular ring in diastole and systole; (c) the nature of the support of the cusp edges; (d) the integrity of cusp tissue around the atrioventricular ring; (e) the mobility and flexibility of the cusps.

Cases with marked insufficiency usually have a deficiency of tissue. This is often most prominent at the septum, where the integrity of the cusp tissue around the ring is in any case interrupted (fig. 5). In extreme cases the cusps may be so short that they do not meet at all, let alone over an area of tissue. This of course may be partly due to ring dilatation.

Attempts to suture the cleft in these circumstances almost inevitably result in a stretched, poorly mobile membrane across the atrioventricular orifice. Extra tissue, perhaps in the form of pericardial autografts, must be supplied, or the valve must be replaced.

Clinical Experience

From January 1962 through January 1963 nine cases with endocardial cushion defects were operated on in the Thoracic Surgical Unit of the University of Cape Town Medical School. All had complete investigations including cardiac catheterization preoperatively. One patient died after surgery. The preoperative diagnosis in this case was that of hypertensive ventricular septal defect, there being
no evidence for mitral insufficiency. The complete form of common atrioventricular canal was found with a ventricular septal defect 1.5 cm. in diameter. A repair was done but the pulmonary hypertension persisted and the patient died suddenly some 18 hours postoperatively and is not included in the table, since no postoperative assessment of mitral valve function could be made. The remaining eight patients have all had repeat clinical and radiologic examinations and catheterizations done 1 year postoperatively.

No case had marked mitral insufficiency although case 2 did have a large left ventricle. The latter case is the only one in fact that demonstrates at all the potential problems and errors catalogued above. This patient was first operated on in 1959. Mitral insufficiency with enlargement of the left ventricle was present preoperatively. There was a tissue deficit at the septum, the gap between the cleft edges at this point being 1 cm. With considerable difficulty the edges were brought together and the whole cleft was stitched. The resulting cusp had impaired mobility and there was residual insufficiency. The atrial septal defect was closed with an Ivalon patch. Both repairs broke down. At the second operation there was some thickening of the cleft edges and less apparent tissue deficit than before. Complete suturing of the cleft produced again a rather immobile, stretched anterior cusp against which the posterior cusp appeared to close reasonably. However, the valve was now definitely stenotic. The anterolateral commissural tissue was partly divided to relieve this stenosis and incompetence inevitably resulted. Although the atrial septal defect is now closed there is still mitral insufficiency with a large left ventricle.

Cases 1 and 3 each had short clefts, one of which was competent and the other not. Each had an additional cleftless and competent orifice to take care of flow, even though there was some narrowing produced by cleft suture. The remaining five cases demonstrate effectively that the principles enunciated above can result in good mitral valve function.

Summary

The mitral valve anatomy of endocardial cushion defects is reviewed and compared with the anatomic requirements for normal mitral valve function. It is shown that very often mitral valve anatomy is such that function should be not at all or only minimally affected. Evidence is quoted that this is indeed so.

The potential harm that may result from indiscriminate suturing of mitral valve clefts and careless repair of the septal defects is described. Principles for the repair of the mitral valve in these cases are enunciated. A small series of cases that have had preoperative and 1-year postoperative assessments of mitral valve function is presented to illustrate some of these points.

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