

Atresia of left atrioventricular orifice

*Anatomical investigation in 62 cases**

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SUMMARY Sixty-two hearts without a patent exit from the left atrium to the ventricular mass have been studied. All had situs solitus and laevocardia. The material consisted of 32 cases with coexistent aortic atresia and 30 cases with patent aortic root. Five hearts with aortic atresia were biventricular with atrioventricular concordance and imperforate left atrioventricular valves, and 27 hearts were univentricular of right ventricular type, with absent left atrioventricular connection. The anatomy of this group was uniform, with extreme hypoplasia of all the left cardiac segments. Among the cases with patent aortic root, five were biventricular, with atrioventricular concordance and imperforate left atrioventricular valves, and 24 had absent left atrioventricular connection, 15 with univentricular heart of right ventricular type and nine with univentricular heart of left ventricular type. The final heart had double inlet univentricle of left ventricular type with an imperforate left atrioventricular valve. In this second group the aorta was larger in cases with discordant ventriculoarterial connection or in those with double outlet from the main ventricular chamber. A normal sized aorta without aortic arch obstruction was observed in nine instances. These are of great interest in terms of surgical anatomy since definitive palliation may be feasible.

“Mitral” atresia has generally been regarded as a congenital cardiac malformation which is beyond complete surgical repair,¹⁻²³ though several ingenious operations have been suggested for its palliation.²⁴⁻²⁵ It is now well established that hearts with “mitral” atresia can coexist with aortic atresia or can have a patent aortic outflow tract. The nature of the “mitral” atresia itself, however, is less well understood. In the case of tricuspid atresia, it has been shown that the lack of a patent exit from the right atrium most frequently results from total absence of the right atrioventricular connection rather than from the presence of an imperforate tricuspid valve.²⁶⁻²⁷ Whether the same morphological features apply to “mitral” atresia remains to be established. Furthermore, conflicting opinions exist of the definition of “mitral” atresia itself, since cases of left atrioventricular atresia with “1-bulbo-ventricular loop” and morphological mitral valve

on the right have been considered as examples of tricuspid atresia on embryological grounds.¹²⁻²⁸ With these considerations in mind we have reviewed the necropsy cases in our cardiopathological collections with atresia of the left atrioventricular orifice. We have then analysed the anomaly with the help of the sequential approach,²⁹⁻³¹ distinguishing those cases with imperforate valves from those with absent left atrioventricular connection. Finally, we have made a special study of those cases with normal aortic pathway, since they are the most suitable candidates for reconstructive surgery.

Material, definitions, and methods

Sixty-two heart specimens were examined in which there was no patent orifice between the left atrium and the ventricular mass (atresia of the left atrioventricular orifice). They were taken from the anatomical collections of the Department of Pathology, Medical School, Padova; Department of Paediatrics, Cardiothoracic Institute, Brompton Hospital, London; and the Thoracic Unit, The Hospital for Sick Children, London.

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The hearts were analysed according to the segmental approach^{29 31} paying particular attention to the ventricular morphology and the associated anomalies.

Absent left atrioventricular connection was considered to be present when there was no connection, either actual or potential, between the left atrium and the ventricular mass. In contrast, if an imperforate membrane was interposed between the two structures, they were considered to be potentially connected, the atrioventricular connection being one of concordance, discordance, or double inlet ventricle, according to the atria and ventricles thus separated (Fig. 1).

When the left atrioventricular connection was

absent, the underlying ventricular chamber was regarded as rudimentary because its inlet was of necessity missing. In contrast, when there was an imperforate valve, the ventricular cavity was considered to be a hypoplastic ventricle rather than a rudimentary chamber, because the inlet was at least potentially present. The rudimentary chamber could be of left or right ventricular type, depending on the morphology of its trabecular zone. When the rudimentary chamber had an arterial exit it was termed an outlet chamber. When it lacked both inlet and outlet portions, it was classified as solely a ventricular trabeculated pouch.

According to the traditional classification of "mitral" atresia, we distinguished two main groups

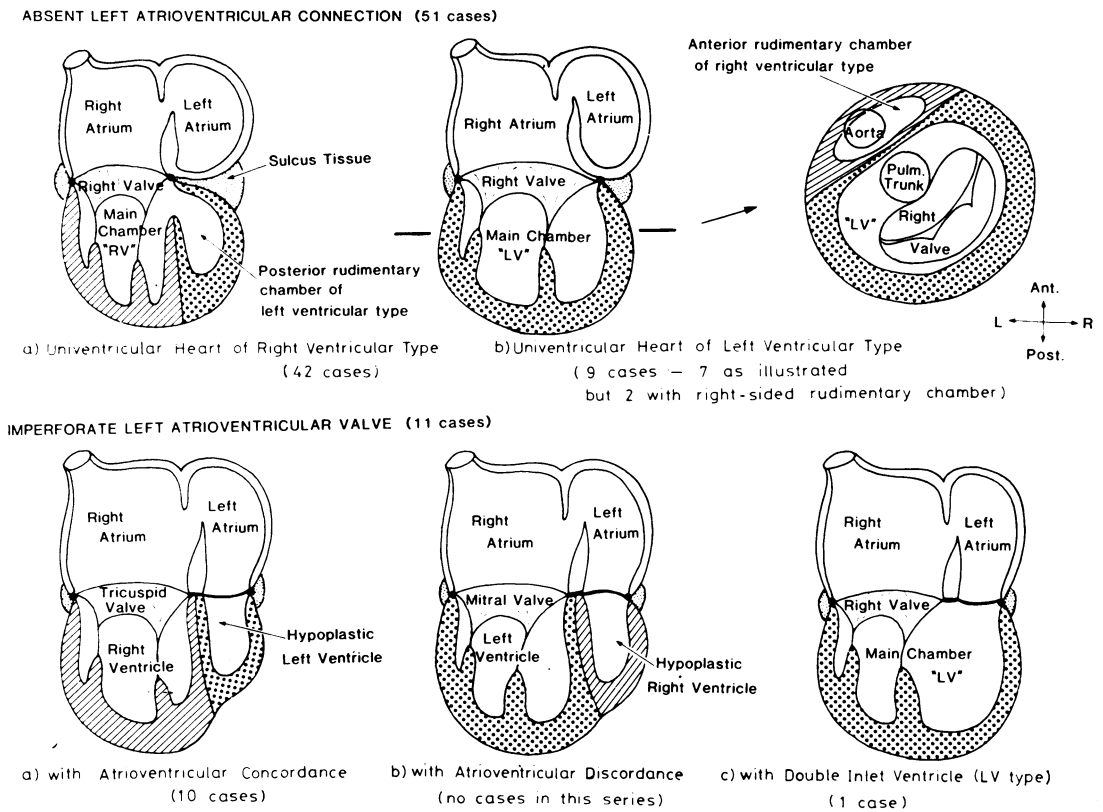


Fig. 1 Diagram illustrating the different types of morphology found at the left atrioventricular junction in the hearts studied. The upper panels show the basic morphology of the 51 cases with absent left connection. Note that sulcus tissue is interposed between left atrium and rudimentary left ventricular chamber in the 42 cases of univentricular heart of right ventricular type. In the nine cases of univentricular heart of left ventricular type, the anterior rudimentary chamber was unrelated to the main chamber, being anterior and left sided in seven and right sided (not illustrated) in two. The bottom panels show imperforate valves in the setting of atrioventricular concordance (10 cases), discordance (included for illustration purposes though no cases were found), and double inlet ventricle (one case of left ventricular type).

of left atrioventricular atresia: (I) With coexistent aortic atresia (32 cases). (II) With patent aortic root (30 cases).

Results (Fig. 1)

(I) LEFT ATRIOVENTRICULAR ATRESIA WITH COEXISTENT AORTIC ATRESIA (32 cases)

All presented with situs solitus and laevocardia. A single pulmonary trunk was present, connected to the right ventricle, with an atretic aorta situated posteriorly and to the right. The ascending aorta and the aortic arch were hypoplastic with a patent ductus arteriosus in all and isthmal coarctation in 19.

According to the nature of the floor of the blinding left atrium, we distinguished two further groups.

(a) *Biventricular hearts with atrioventricular concordance and imperforate left atrioventricular valve* (five cases, four boys and one girl, average age 4 days)

The annulus of the imperforate valve was very small in each. A subvalvular tensor apparatus was found

in four of these. A hypoplastic left ventricle was always present, being posterior and left-sided and was consistently reduced to a slit-like structure.

Further cardiac anomalies were: intact atrial septum (one case); atrial septal defect, fossa ovalis type (one case); total anomalous pulmonary venous drainage to superior vena cava and persistent left superior vena cava (one case).

(b) *Univentricular hearts of right ventricular type with absent left atrioventricular connection*

(27 cases, 14 boys and 13 girls, average age 7 days).

In each case a rudimentary chamber of left ventricular type was identified posteriorly and on the left. The chamber was a slit-like trabecular pouch in 24 cases. In three hearts, in which there was also an apical defect or multiple ventricular septal defects, the rudimentary chamber was slightly larger, but unequivocally small.

In this subgroup coexisting anomalies were: anomalous pulmonary venous return partial to the superior vena cava (one case) and total to persistent left superior vena cava (one case); left cor triatriatum

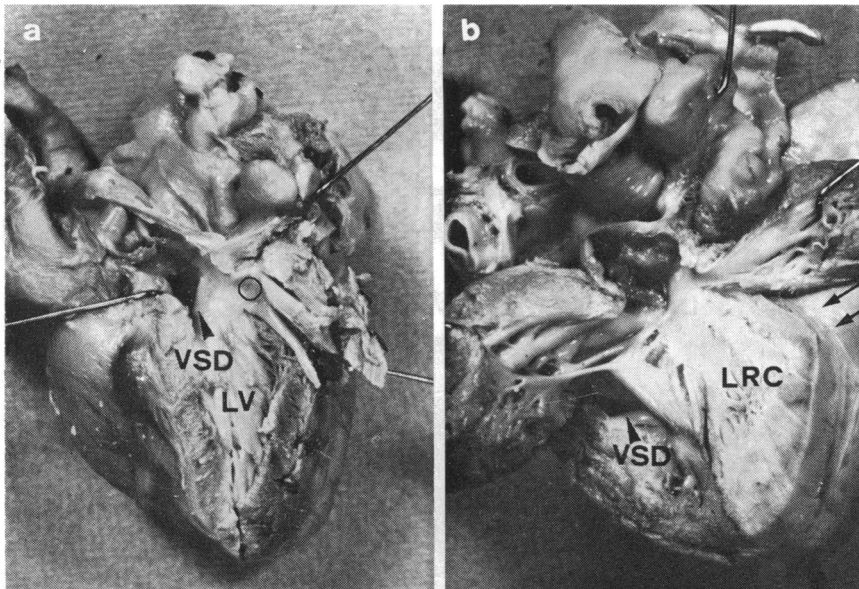


Fig. 2 (a) *Biventricular heart with concordant atrioventricular connection and imperforate left atrioventricular valve, concordant ventriculoarterial connection and patent aortic root. The circle indicates the small imperforate membrane separating the left atrium from the hypoplastic left ventricle (LV). The ventricular septal defect (VSD) is small and subaortic.* (b) *Univentricular heart of right ventricular type with absent left atrioventricular connection and concordant ventriculoarterial connection. The rudimentary chamber was of left ventricular type (LRC) and the posterior component of the ventricular septum is present reaching the crux cordis and the descending coronary artery (arrows). The ventricular septal defect (VSD) is muscular and anterior.*

(two cases); intact atrial septum (four cases), one of which had agenesis of the coronary sinus and persistent left superior vena cava; atrial septal defect of fossa ovalis type (six cases), and finally anomalous origin of the right subclavian artery (one case).

(II) LEFT ATRIOVENTRICULAR ATRESIA WITH PATENT AORTIC ROOT (30 cases)

These cases all presented with situs solitus and laevocardia. According to the atrioventricular connection and the morphology of the ventricular mass, they were subdivided into groups of:

(a) Biventricular hearts with atrioventricular concordance and imperforate left atrioventricular valve (five cases, two boys and three girls, ages varying from 8 to 37 days, average 15)

The imperforate orifice consisted of a very small membrane in four (Fig. 2a) and of a better formed membrane with a discrete tensor apparatus in one. The left ventricle, which was situated posteriorly and on the left, was hypoplastic in all.

Three cases presented with a concordant ventriculoarterial connection, one with intact ventricular septum and two with multiple ventricular septal defects, and the aorta was severely hypoplas-

tic in all three. The case with intact septum was particularly unusual since, though there was no way into the tiny left ventricular chamber either through the mitral orifice or through a ventricular septal defect, there was a patent aortic root. Two cases had double outlet right ventricle, one with subaortic ventricular septal defect and one with intact ventricular septum; the size of the aorta in these two was about half the size of the pulmonary trunk.

All patients showed coarctation and/or tubular hypoplasia of the aortic isthmus with patent ductus arteriosus.

Further associated anomalies were: left cor triatriatum (one case); persistent left superior vena cava draining into coronary sinus (one case); bicuspid aortic valve with discrete subaortic stenosis (one case); intact atrial septum with left atrial fibroelastosis (one case); total anomalous pulmonary venous drainage into superior vena cava (one case).

(b) Univentricular heart of right ventricular type with absent left atrioventricular connection (15 cases, seven boys and eight girls, ages varying from 1 to 135 days, average 30)

The rudimentary chamber was always situated

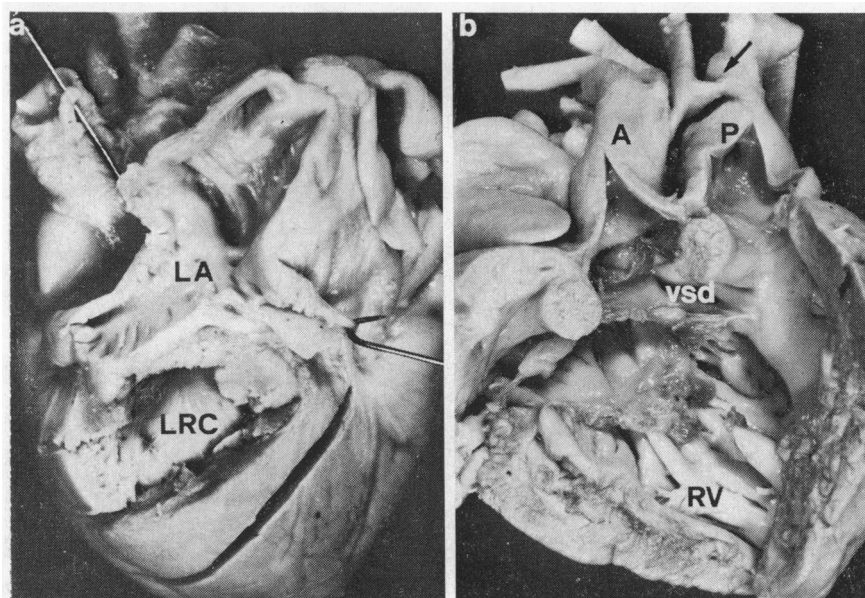


Fig. 3 Univentricular heart of right ventricular type with absent left atrioventricular connection and double outlet right ventricle. (a) Left side view: note the absence of a connection between the left atrium (LA) and the underlying left ventricular cavity, which is a slit-like rudimentary chamber (LRC). (b) Both the great arteries arise from the right ventricle (RV), the aorta (A) being on the right and posterior to the pulmonary trunk (P). The arrow indicates tubular hypoplasia of the aortic arch. The ventricular septal defect (vsd) is subaortic.

posteriorly and on the left. The posterior component of the ventricular septum in all cases reached the crux cordis (Fig. 2b). The ventriculoarterial connection was concordant in six cases and discordant in one, the left ventricular rudimentary chamber in all these cases being a hypoplastic outlet chamber. In contrast, in seven patients with double outlet from right ventricular chamber and in the one with single outlet and pulmonary atresia, the rudimentary chamber was a slit-like cavity with no arterial outlet and was regarded as a left ventricular trabecular pouch (Fig. 3). All the six patients with concordant ventriculoarterial connection had septal defects: one multiple, two perimembranous, and three muscular. The case with discordant ventriculoarterial connection had a restrictive subpulmonary defect (Fig. 4). Within the group with double outlet, three had an apparently intact ventricular septum, three had a perimembranous subaortic defect, and one a subpulmonary septal defect. The case with single outlet and pulmonary atresia had a restrictive subaortic defect.

The size of the aorta was less than one-third the size of the pulmonary trunk in the cases with concordant ventriculoarterial connection, but only one of these patients had an unobstructed aortic arch. The patients with a discordant ventriculoarterial connection and with single outlet had a large aorta and aortic arch. The seven cases with double outlet

had an ascending aorta of good size (about half the diameter of the pulmonary trunk) but three had coarctation and one tubular hypoplasia of the aortic arch.

Other associated anomalies were: atrial septal defect, fossa ovalis type (five cases); restrictive foramen ovale (two cases); intact atrial septum (one case); persistent left superior vena cava draining to the coronary sinus (four cases); partial anomalous pulmonary venous drainage in superior vena cava (one case); bicuspid aortic valve (one case); bicuspid pulmonary valve (one case); persistent ductus arteriosus (nine cases).

(c) *Univentricular hearts of left ventricular type with absent left atrioventricular connection*

(nine cases, five boys and four girls, ages varying from 10 to 215 days, average 95)

In these patients the rudimentary chamber was of right ventricular morphology and was situated anteriorly and to the right in two cases and anteriorly and to the left in seven (Fig. 5).

The right atrioventricular valve, interposed between the right atrium and the main left ventricular chamber, was morphologically a mitral valve. The trabecular septum, which separated the main from the rudimentary chamber never extended to the crux cordis. The outlet foramen between the main chamber and the rudimentary chamber was

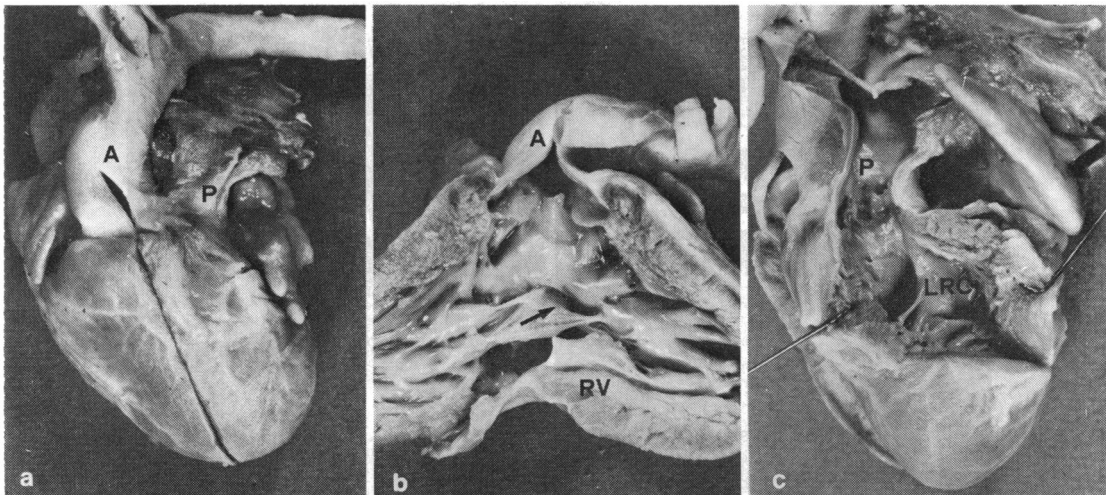


Fig. 4 Univentricular heart of right ventricular type with absent left atrioventricular connection and discordant ventriculoarterial connection. (a) Anterior view showing the right-sided large aorta (A). Note the unobstructed aortic arch. (b) The aorta takes origin from the main chamber of right ventricular type (RV); the arrow indicates the restrictive ventricular septal defect. (c) The pulmonary artery (P) takes origin mainly from the rudimentary chamber of left ventricular type (LRC).

restrictive in three cases. The ventriculoarterial connection was discordant in seven, double outlet from the left ventricular chamber in one, and single outlet with pulmonary atresia in one. The size of the rudimentary chamber was in each case very small. The aorta, which was anterior in each patient, was of normal size in six, all also having a normal aortic arch. The other three patients had restrictive outlet foramen with hypoplastic ascending aorta, preductal coarctation, and patent ductus arteriosus.

Further associated anomalies were: patent ductus arteriosus in the case with pulmonary atresia; atrial septal defect of fossa ovalis type (one case); restrictive foramen ovale (two cases); absence of the coronary sinus (one case); stenotic bicuspid pulmonary valve (one case).

(d) Univentricular heart of left ventricular type with double inlet and imperforate left atrioventricular valve (one girl, 1 month old)

In this case the left atrium was separated from the main ventricular chamber by an imperforate membrane, the right atrium also being connected to the main ventricular chamber which had left ventricular morphology. The rudimentary chamber

of right ventricular type was anterior and to the right and was separated from the main chamber by an anterior trabecular septum which did not extend to the crux. The outlet foramen was not restrictive. The rudimentary chamber gave origin to the aorta which was smaller than the pulmonary artery. There was mild isthmal coarctation and the ductus was patent.

Discussion

DEFINITION, NOMENCLATURE, AND NOSOLOGICAL CONSIDERATIONS

The hearts described in this investigation were unified by atresia of the left atrioventricular orifice. Because there was situs solitus in each case, this lesion blocked egress of the pulmonary venous return from the left-sided, morphologically left atrium to the ventricles. Included in this series are not only cases in which a morphologically tricuspid valve connects the right atrium to a morphologically right ventricle, but also cases in which a morphologically mitral valve connected the right atrium to a morphologically left ventricle. The latter cases have previously been described by Tandon and Edwards²⁸ as tricuspid atresia, despite the fact

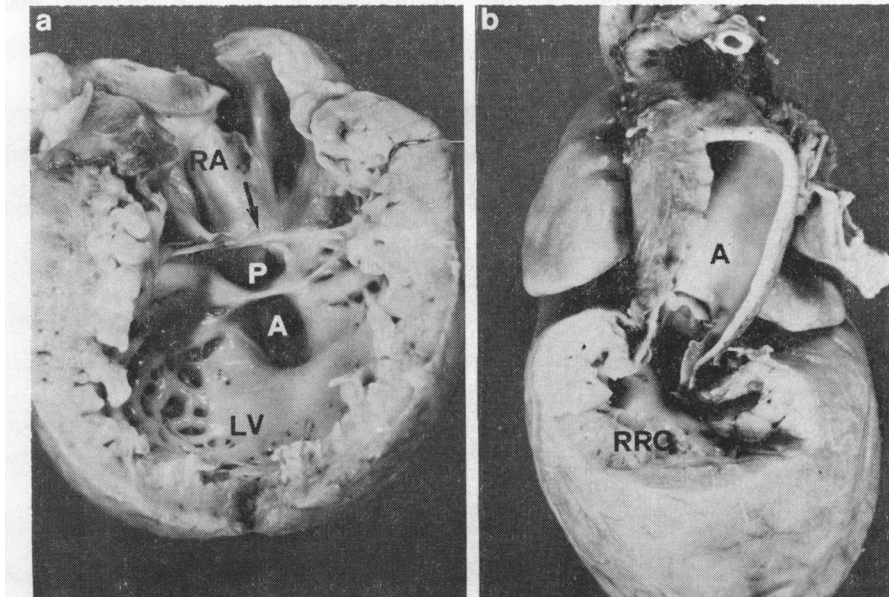


Fig. 5 Univentricular heart of left ventricular type with absent left atrioventricular connection and double outlet left ventricle. (a) View from the apex showing a mitral valve (arrow) connecting the right atrium (RA) with the main chamber of left ventricular type (LV). Both the aorta (A) and the pulmonary artery (P) arise from the left ventricle. (b) Left lateral view which shows the small rudimentary chamber of right ventricular type (RRC) and the ascending aorta.

that the atresia was on the left. The rationale for the description of tricuspid atresia was that the atretic valve would have been a tricuspid valve, had it been present. For the same reason, Moreno *et al.*¹² excluded these cases from their review of mitral atresia. Using the criteria of these authors, atresia of the right atrioventricular orifice, when a morphologically tricuspid valve connects a left atrium to a morphologically right ventricle, could then correctly be described as mitral atresia. However, Vlad³² has included an example of this last anomaly as tricuspid atresia. We believe that the most important feature of these hearts from clinical and surgical standpoints is the nature of the atrium which has no normal exit. Thus, in all the cases we have described, it is the left atrium which lacks a direct opening to the ventricles, irrespective of whether the atretic valve, had it been formed, would have been morphologically mitral or tricuspid. To avoid the confusion and discrepancies in nomenclature indicated above, we therefore prefer to distinguish between atresia of the right and left atrioventricular orifices rather than between "tricuspid" and "mitral" atresia.³⁰

According to our findings, the morphological substrate of the atresia between the left atrium and the ventricular mass is in the majority of cases an absent atrioventricular connection, with only few cases showing an imperforate valve. In the former condition there is absence of the entire left atrioventricular valve tension apparatus, in addition to absence of the atrioventricular connection. In the latter situation, hypoplastic elements of valve tension apparatus are observed and a potential connection exists between the left atrium and the ventricular mass. It follows from this morphology that most of the hearts with atresia of the left atrioventricular orifice present with only one ventricular chamber possessing an atrioventricular connection. Since it is our convention to group together as univentricular hearts those in which the atrioventricular connection or connections are committed to only one ventricular chamber, the hearts presently studied with absent left atrioventricular connection fall into our category of univentricular hearts. We contest the usual definition by which cases of atrioventricular orifice atresia have to be excluded from the category of single ventricle or univentricular hearts.^{33 34} We prefer to categorise a ventricular chamber as rudimentary if it does not receive an atrioventricular connection. It does not matter whether the main ventricular chamber has an atrioventricular connection via two valves, a common valve, or a single right or left valve, since this does not affect the morphology of the other chamber.

An imperforate left atrioventricular valve was

associated in all but one of our cases with a potentially concordant atrioventricular connection, the hearts thus being biventricular. In the outstanding case both the imperforate left-sided valve and the patent right-sided valve connected the left and right atria with the main left ventricular chamber, as seen in double inlet to a univentricular heart of left ventricular type, when both valves are patent. An imperforate left atrioventricular valve with a discordant connection (congenitally corrected transposition) has not been thus far described to our knowledge.

Univentricular hearts with an atretic left atrioventricular orifice were found to exhibit the same variability in ventricular morphology as was previously found in univentricular hearts with double inlet.^{34 35} Thus, we found main chambers of both right and left ventricular type connected to the right atrium, and expect in future to encounter cases with sole chamber of indeterminate morphology. The cases we studied with main chamber of right ventricular type had rudimentary chambers with left ventricular features. The cases with main chamber of left ventricular type had rudimentary chambers with right ventricular features. The left ventricular rudimentary chambers were always posterior and to the left, while the right ventricular rudimentary chambers were always anterior, in two cases being right sided and in seven being left sided. The ventricular septum which separated the main from the rudimentary chamber had a different orientation in the two groups. In the univentricular hearts of right ventricular type, the ventricular septum always extended to the crux cordis, while in the univentricular hearts of left ventricular type the septum was always anterior and never extended to the crux. This different orientation is hardly surprising since the rudimentary chamber of left ventricular type is derived from the ventricular trabeculated pouch which grows from the inlet part of the ventricular loop (embryonic primitive ventricle) while the rudimentary chamber of right ventricular type is derived from the ventricular trabecular pouch which grows from the outlet part of the loop (embryonic bulbus).

CLINICAL AND SURGICAL IMPLICATIONS

The feature determining survival among patients with atresia of the left atrioventricular orifice is not the atresia itself but the presence of a patent aortic root and/or isthmal hypoplasia. Cases with co-existent "aortic and mitral" atresia had uniform anatomy, with extreme hypoplasia of all the left cardiac segments. This accounts for the bad prognosis and early death.²³ In this condition the

systemic circulation depends always upon the patency of the ductus. They are comparable with cases of aortic atresia, intact ventricular septum, and hypoplasia of the mitral valve and left atrium, but left fibroelastosis is significantly missing in the cases with atretic left atrioventricular orifice. It is interesting that we were able to identify two ventricular chambers in all hearts, albeit after careful dissection, often guided by the coronary artery anatomy. Accordingly, we doubt the existence of cases of "mitroaortic" atresia with absent left ventricular cavity, as reported by some authors.³⁶

Cases with patent aortic root have a better prognosis even though other severe cardiac anomalies are commonly associated. The term "isolated mitral atresia" which has been used to describe this condition does not seem appropriate, since it is in fact never isolated. The interatrial shunt is frequently obstructed,¹¹ but, if not, survival in these patients is mainly dependent upon aortic blood flow and the status of the aortic arch. Our observations on the size of the aorta suggest that the aortic blood flow was inadequate in cases with ventriculoarterial concordance. In these hearts the aorta, which arose from the left ventricular outlet chamber or from the diminutive left ventricle, was always hypoplastic and there was coarctation and/or tubular hypoplasia of the aortic arch. We suggest this is because the ventricular septal defect in cases with a concordant ventriculoarterial connection was always restrictive. The calibre of the aorta arising from the left ventricular cavity is essentially dependent on the size of the defect and the amount of the right to the left shunt at this level.

The aorta was larger in cases with a discordant ventriculoarterial connection or in those with double outlet from the main chamber. In particular, an aorta near normal in size without isthmal obstruction was seen in two patients with double outlet right ventricle, in one with discordant ventriculoarterial connection and univentricular heart of right ventricular type, and in six with univentricular hearts of left ventricular type. In this condition the aorta had a favourable exit from the heart since it took origin from the main chamber or from the rudimentary chamber of right ventricular type without restriction of the outlet foramen. The three cases with univentricular heart of left ventricular type, however, which did have aortic arch obstruction, also had a restrictive outlet foramen.

One of our cases of univentricular heart with right ventricular main chamber and left ventricular outlet chamber had "transposition" of the great arteries, to our knowledge the first time that this ventriculoarterial connection has been found in a univentricular heart of right ventricular type. A similar ven-

triculoarterial connection has been recently reported by Ostermeyer *et al.*,²⁵ but in their case an imperforate left atrioventricular valve was present, so that within our categorisation their heart was biventricular. Indeed, they have misinterpreted our concept when they state that we would categorise their heart as univentricular.

The distinction of an absent connection from an imperforate valve is the important feature, and is of clinical relevance since an imperforate atrioventricular valve may be recognised angiographically with selective injection into the hypoplastic ventricular chamber.^{8,9}

The patients with a normal aorta are the only ones in which a definitive surgical palliation might be reasonably considered. Atrial septectomy or septostomy (either balloon or blade), for the relief of obstructed interatrial shunt, should be combined with a systemic-to-pulmonary artery anastomosis in cases with pulmonary outflow stenosis, or with pulmonary banding when the pulmonary blood flow is excessive.

The morphology of either the hypoplastic left ventricles or the tiny rudimentary chambers suggests that these ventricular cavities could never be used as ventricles for propelling blood flow to a great artery even should definitive reconstruction of the atrioventricular connection be possible. Moreover, the presence or absence of a left atrioventricular connection did not affect the size of the ventricular cavity, whether it was a left ventricle or a rudimentary chamber. These were always diminutive in size. Indeed the imperforate atrioventricular valve was never so large that the left atrioventricular connection could be restored by a prosthetic valve. Possible operations in theoretical cases with a left ventricle or rudimentary chamber of adequate size have been recently postulated, as have "reversed Fontan" operation in which systemic venous return is directed to the pulmonary artery and pulmonary venous return to the main ventricular chamber. To our knowledge, however, successful operations of these types have yet to be reported.

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