Atresia of the right atrioventricular orifice

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The results are reported of a study of 83 necropsied hearts with atresia of the right atrioventricular orifice. It is emphasised that right atrial or atrioventricular orificial atresia is a better term to describe this anomaly than 'tricuspid atresia'. Use of the latter term can be confusing when the morphologically tricuspid valve is located beneath the left atrium. It is accepted that the definition employed may include cases in which the mitral valve may be atretic, blocking normal exit from the right atrium, but it is argued that such cases would present clinically as 'tricuspid atresia' and therefore are correctly designated as right atrial orificial atresia. The results show that the majority of hearts with right atrial orificial atresia have the ventricular morphology of primitive ventricle, most with, but a few without an outlet chamber. However, in a minority of hearts an imperforate membrane interposes between the right atrium and a formed but hypoplastic right ventricle. In two of the hearts, the imperforate membrane showed features of Ebstein's malformation. The hearts could be further subdivided according to the ventriculocardiacal connection. Most had normally connected arteries (66 of 83), and all but 2 also had normal relations between the arteries; in these 2 hearts there was 'anatomically corrected malposition'. Twelve hearts showed transposition, one had double-outlet outlet chamber, and another persistent truncus arteriosus. The remaining 3 hearts, all without outlet chamber, had by definition a double outlet connection. A segmental approach provides the best way of classifying this anomaly, and an embryological explanation is offered for the variations in anatomy observed.

The classification of tricuspid atresia introduced by Edwards and Burchell (1949) has proved its value in clinical, surgical, and pathological settings. However, recent modifications of the original classification have perhaps detracted from its value. Using the modification suggested by Keith et al. (1958), atresia of either the right or left atrioventricular valves can correctly be described as tricuspid atresia. Definition of 'transposition' as applied to this classification varies considerably and is a potential source of further confusion. Thus, using the term 'transposition' to describe any anteriorly situated aorta, Tandon and Edwards (1974) described under the heading of 'tricuspid atresia with l-transposition' first, hearts with right atrioventricular valve atresia in which the aorta arose from the left ventricle and secondly, hearts with left atrioventricular valve atresia in which the aorta arose from the right ventricle.

With the advent of radical palliative surgery for tricuspid atresia (Fontan and Baudet, 1971), it is desirable to have a system of nomenclature which avoids these discrepancies. Any successful nomenclature should permit accurate description of hearts in all aspects of their anatomy, in particular with regard to the site of atrial orificial atresia and to the ventricular morphology and ventriculocardiacal connections. The segmental approach promoted by Van Praagh and his colleagues (Van Praagh et al., 1964; Melhuish and Van Praagh, 1968; Van Praagh, 1972) is ideally suited for this purpose. We have therefore analysed a series of specimens with right atrial orificial atresia using this approach.

Materials and methods

The 83 hearts studied were all those with atresia of the right atrioventricular orifice in the pathological collections of the Brompton Hospital, London; Royal Liverpool Children's Hospital; Grimsby General Hospital; Wilhelmina Gasthuis, Amsterdam; Sint Antonius Ziekenhuis, Utrecht; and selected specimens from the Department of Anatomy, University of Leiden. In all instances the
hearts were studied so as to elucidate their segmental arrangement, paying particular attention to the ventricular morphology, and the nature of associated anomalies. Twelve of the hearts were also studied by serial histological sectioning of the atrioventricular junctions primarily in order to ascertain the disposition of the conducting tissue (J. L. Wilkinson, A. Smith, A. E. Becker, and R. H. Anderson, unpublished observations). Certain of these histological findings are pertinent to the present investigation, and will be discussed where relevant.

Definitions and terms

(1) ATRIAL SITUS
Situs solitus is present when the morphologically right atrium is right sided and the left atrium is left sided (Shinebourne et al., 1976). The other varieties of atrial situs are situs inversus and situs ambiguus.

(2) VENTRICULAR MORPHOLOGY
The terms right ventricle and left ventricle will only be used in this report when each ventricular chamber is in potential communication with a separate atrial chamber, and each ventricular chamber contains the complete tension apparatus of the connecting atrioventricular valve (Fig. 1).

In the situation in which a single atrioventricular valve is present between the left atrium and a ventricular chamber, and the blind-ending right atrium is also in potential communication with the chamber receiving the single atrioventricular valve, then the ventricular morphology is considered to be a variant of primitive (single or common) ventricle (Macartney et al., 1976; Shinebourne et al., 1976). In this situation the chamber receiving the single atrioventricular valve is termed the main chamber. If a smaller anterior chamber is also present it is termed the outlet chamber (Fig. 2). The foramen...
between the main and outlet chambers is described as the bulboventricular foramen, remembering that if one great artery arises from the main chamber the foramen should strictly be termed the secondary bulboventricular foramen. (Compare closure of the interventricular defect by Van Mierop et al. (1963).)

When a single ventricular chamber receives a single atrioventricular valve, supports both great arteries, and also is in potential communication with the blind-ending right atrium, there being no evidence of an anterior outlet chamber, then the anomaly is described as primitive (single or common) ventricle without outlet chamber (Fig. 3). The single atrioventricular valve present in the hearts to be described is termed the left atrioventricular valve. Its morphology will be described where pertinent. Since the blind-ending right atrium does not always have a fibrous membrane interposing between it and a ventricular chamber, the situation will be described as right atrial orificial atresia rather than as valve atresia. If a membrane is present, then its morphology will be described.

3) VENTRICULOARTERIAL CONNECTIONS

(Fig. 4)

Normal connections describe the situation in which the aorta arises from the left ventricle or from the main chamber, and the pulmonary artery from the right ventricle or from the outlet chamber. Usually with normal connections the aortic valve is posterior and to the right of the pulmonary valve. This is described as normal relations (Fig. 4A). Occasionally normal connections exist with the aortic valve in anterior, left-sided position (Fig. 4B). This is described as normal connections with l-malposition or 'anatomically corrected malposition'. Transposition is present only when the aorta arises from the right ventricle (or from an outlet chamber) and the pulmonary artery from the left ventricle (or from the main chamber). The prefixes d- and l- refer to the position of the aortic valve relative to the pulmonary valve whether describing malposition or transposition (Fig. 4C and D). When more than 1½ great arteries arise from the same chamber, the connection is described as double-outlet main chamber or double-outlet outlet chamber (Fig. 4E); the great arteries in double-outlet chambers are described in terms of d- or l-malposition. When a single arterial trunk leaves the base of the heart and when this single artery gives rise to the aorta, one or both pulmonary arteries, and the coronary arteries, it is described as a persistent truncus arteriosus (Fig. 4F). Other variants of single trunk are single aortic trunk with pulmonary atresia and single pulmonary trunk with aortic atresia.

4) VENTRICULAR MUSCLE BUNDLES

In the hearts studied the muscular septum interposing between the aortic and pulmonary valves is described as the infundibular septum. Previously this structure has been termed the conus septum (Anderson et al., 1974b). If a muscular fold separates the posterior semilunar valve from the atrioventricular valve it is termed the ventriculo-infundibular fold. This previously has been termed the conoventricular flange (Anderson et al., 1974b).

Results

A. ATRIAL ANATOMY

All the hearts studied had situs solitus. The right atrium, therefore, received the systemic venous return. The only exit from the right atrial chamber was through an interatrial communication. In 77 hearts, the communication was either a secundum atrial septal defect or a widely patent foramen ovale.

In 6 hearts the defect was of the ostium primum variety, in each case with a cleft in the left atrioventricular valve (Fig. 5). A further significant
Right AV valve atresia

Ventriculoarterial connections

A) Normal relations—58 cases
   (4 with Pulmonary atresia)
B) L-malposition—2 cases

Normal connections

C) d-transposition—10 cases
D) l-transposition—2 cases

Transposition

E) Double outlet
   (outlet chamber)
   1 case
F) Single arterial trunk
   (persistent truncus arteriosus)
   1 case

Fig. 4 Diagram illustrating ventriculo-
arterial connections in 74 cases of primitive
ventricle with outlet chamber and right
atrial orificial atresia, and the number of
specimens studied in each category.

Fig. 5 Photographs of a heart with primitive ventricle and outlet chamber and right atrial orificial atresia, an ostium
primum defect, and a cleft mitral valve. (A) View from posterior aspect of main chamber. The free lower edge of the
septum primum (SP) is seen, with an ostium primum (OP) defect beneath it. The mitral valve (MV) has a cleft
(arrowed) in its anterior leaflet, and the valve orifice extends into the right atrium. The cut seen is through the right
lateral wall of the heart. (B) Magnified view of cleft in anterior mitral leaflet.
feature of right atrial anatomy in approximately a
third of the hearts studied was persistence of
portions of the embryonic venous valves. In 2
instances the valves were almost complete and
partitioned the auricular part of the chamber from
the sinus venarum (Gerlis and Anderson, 1976).
In one heart, Fontan repair had been attempted,
and the Eustachian valve formed a veil across the
prosthetic inferior vena caval valve which had been
inserted, separating it from the atrial insertion of the
conduit. Discrete ‘dimples’ in the floor of the right
atrium were observed in 36 of the hearts studied.
The left atrium in all hearts received the pulmonary
venous return and, via the interatrial communication,
the systemic venous return.

B. VENTRICULAR MORPHOLOGY
In 6 hearts formed right and left ventricles were
identified (Fig. 1). An imperforate membrane was
present in the floor of the right atrium separating it
from the formed right ventricle. On its ventricular
aspect the membrane was attached to hypoplastic
valvar tension apparatus. In 4 of the 6, the right
ventricle was small and hypoplastic and the infundibi-
bulum was also blind ending. These hearts were,
therefore, examples of pulmonary atresia with
intact septum. In one the cavity extended as
multiple sinusoids into the septum, which bulged
into the aortic outflow tract. In the other 2 hearts,
the imperforate membrane between right atrium
and right ventricle was displaced distally to the

Fig. 6 Photographs of a heart with
formed right and left ventricles and an
imperforate Ebstein’s anomaly. (A) An
anterior view of the heart. The arteries
(Ao, PA) are normally connected and
normally related. (B) The right side of
the heart after removal of its right
posterolateral wall. The right atrium
(RA) leads to the right ventricular
sinus (RVS) but egress of blood is
blocked by an imperforate tricuspid
valve. The ventricular origin of the
valve (arrows) is well below the annulus
fibrosus (AF) indicating an Ebstein
anomaly. (C) The opened infundibulum
(Inf). Blood reaches the pulmonary
artery (PA) through a large ventricular
septal defect (VSD), and the
imperforate tricuspid valve is seen
through the defect. Note also the
prominent trabecula septomarginalis
forming a partial septum between the
infundibulum and sinus of the right
ventricle. This structure could also be
considered a remnant of the bulbo-
ventricular septum. (D) The left-sided
chamber is a morphological left ventricle
(LV). The defect from this chamber to
the infundibulum is now bounded by the
interventricular septum and the
infundibular septum (IS) not by the
right bulboventricular ridge as in the
hearts with primitive ventricle (vide
infra). LA, left atrium; MV, mitral
valve; Ao, aorta.
junction of ventricular inflow and outflow portions. They were, therefore, examples of imperforate Ebstein's anomaly. One of these has been described elsewhere (Gerlis and Anderson, 1976); the other (Fig. 6) had a large ventricular septal defect between the left ventricle and the right ventricular infundibulum, which supported a good sized pulmonary artery.

In 77 hearts the blind ending right atrium was not in potential communication with an anterior ventricle, but with the same ventricular chamber as received the left atrioventricular valve (Fig. 7 and 8). All these hearts were, therefore, examples of primitive (single or common) ventricle. In one heart (Fig. 7) a fibrous membrane at the site of the dimple formed a discrete aneurysm protruding from the floor of the right atrium into the main chamber. In 36 of the remaining hearts the right atrial 'dimple' overlaid the central fibrous body, and was again the potential communication with the main chamber. In 34 hearts, discrete 'dimples' were not observed, but dissection revealed that the right atrial floor was in potential communication only with the main chamber (Fig. 8). In a further 6 hearts, ostium primum defects existed through which the right atrium was in direct communica-

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**Fig. 7** Photographs of a specimen of primitive ventricle with outlet chamber, right atrial orificial atresia, and with an aneurysmal 'dimple'. (A) Anterior view of the heart (OC, outlet chamber; MC, main chamber; Ao, aorta; PA, pulmonary artery). (B) Right atrial anatomy. The dimple is large and aneurysmal (An) (FO, foramen ovale; CS, coronary sinus). (C) Left atrium communicating with main chamber through a normal mitral valve (MV) (LAA, left auricular appendage; APM, PPM, anterior and posterior papillary muscles). (D) After retraction of the mitral valve, the aneurysm is seen pointing into the large cleft between the well formed posterior ridge (PR) and the bulboventricular septum. We believe this cleft to represent the unexpanded right ventricular inflow tract (BVF, bulboventricular foramen).
tion with the main chamber (Fig. 5). The histo-
logical studies in 12 hearts confirmed these atrio-
ventricular relations, and showed that the atrioven-
tricular incisura always interposed between the
right atrial floor and the outlet chamber.

In 74 of the 77 hearts the ventricular morphology
was that of primitive ventricle with outlet chamber.
In the remaining 3 hearts there was only one ven-
tricular chamber, which received the left atrioven-
tricular valve and gave rise to both great arteries
(Fig. 9). In primitive ventricle without outlet

chamber the ventriculoarterial connection must by
definition be one of double outlet (Shinebourne
et al., 1976). There was d-malposition in two of
these hearts and l-malposition in the third. When an
outlet chamber was present, it was anterior and to
the right in 71 specimens and to the left in 3. The
outlet chamber morphology varied according to the
ventriculoarterial connection. With normal ventri-
culoarterial connection, the outlet chambers had
long infundibular segments and smaller apically
 trabeculated portions (Fig. 2, 10). In contrast, when

Fig. 8. Photographs of a specimen of primitive ventricle with outlet chamber, right atrial
 orificial atresia, and transposed great arteries. The general
 view of the heart is seen in (D) (RAA-
 right auricular appendage). (A) Detail of the blind-
 ending right atrium. Note the prominent
 right venous valve (RVV). (B) Detail of the anterior aspect of the opened outlet
 chamber. Note that the
 infundibular septum is short so that the
 bulboventricular
 foramen opens directly beneath the aortic valve
 (compare Fig. 8B,
 10B). (C) Detail of a right oblique view of the heart. The right
coronary artery (RCA)
 runs between the right
 atrium and the outlet
 chamber, and its
descending branch
 (RDA) delimits the
 outlet chamber. The
 chamber is not in
 contact with the right
 atrium. (E) Main
 chamber giving rise to
 the pulmonary artery.
 Note the prominent
 posterior ridge and
 mitral-pulmonary
 valvar continuity
 (between arrows).
 Other abbreviations as
 before.
transposition was present the infundibular segment was much shorter and the trabeculated zone more prominent (Fig. 8).

The trabeculations of the main chamber resembled those of the normal left ventricle in all cases, but were frequently much coarser than normal (Fig. 10). There were several other differences between the main chamber and a normal left ventricle. The septal surface was much smaller, was orientated in the frontal plane, and was frequently trabeculated. The bulboventricular defect was separated from the area of the central fibrous body by direct fusion of the infundibular septum with the bulboventricular septum. The aortic valve in hearts with normally connected arteries was, therefore, separated from the septal valve by a muscular bar, unlike the situation seen in isolated ventricular septal defect with normally connected arteries (compare Fig. 11A and B). Another unusual feature of the main chamber was the presence of a prominent posterior trabeculation. In some hearts this ridge was sufficiently prominent to divide the chamber into large left and smaller right-sided portions (Fig. 10). In all but two hearts the valve entering the main chamber had the morphological characteristics of a mitral valve (Fig. 7, 8). In these two hearts, both with left-sided outlet chambers, there were morphologically tricuspid valves between the left atrium and the main chamber; in both hearts the outlet chamber was larger with a prominent trabeculated pouch and the bulboventricular septum was straddled by the left atrioventricular valve (see Fig. 13). In the other

![Fig. 9 Photographs of a specimen of primitive ventricle, without outlet chamber, and right atrial orificial atresia. (A) The general view of the heart. The auricles (RAA, LAA) are juxtaposed to the left of the great arteries, the aorta (Ao) being anterior and to the right of the pulmonary artery (PA). (B) The blind-ending right atrium (RA). (C) The anterior-aorta (Ao) arises from the sole ventricular chamber (VC). The cavity of the ventricle is spanned by a free running strand which represents the trabecula septomarginalis (TSM). These relations are better illustrated in (D), taken from the apex of the ventricle. The single chamber receives the left atrioventricular valve (LAVV) and gives rise to both great arteries (Ao, PA), which are separated by the infundibular septum. The ventriculoarterial connection is one of double outlet ventricle.](http://heart.bmj.com/first-published-as-10.1136/hrt.39.4.414-on-1-april-1977/10.1136/hrt.39.4.414-on-1-april-1977)
Fig. 10 Further views of the case of primitive ventricle with outlet chamber, right atrial orificial atresia, and aneurysmal 'dimple' illustrated in Fig. 7. (A) The relation of the bulboventricular foramen to the posterior ridge. The inferior edge of the infundibular septum (IS) and the bulboventricular septum (BVS) separate the dimple from the outlet chamber. (B) The anterior aspect of the outlet chamber. It differs in morphology from that seen in Fig. 8, by virtue of the deep infundibulum (Inf) beneath the pulmonary valve. It possessed an extensive trabeculated apical pouch (TrP). However, it does not contain tension apparatus and does not receive the aneurysmal dimple (see Fig. 7).

Fig. 11 Photographs illustrating the differences between an isolated ventricular septal defect (VSD) in a heart with atrioventricular concordance (A), and a secondary bulboventricular foramen (BVF) in a heart with primitive ventricle with outlet chamber and right atrial orificial atresia (B). (A) Viewed from the left ventricle (LV); (B) from the main chamber (MC). Note that in the isolated defect, the roof is the aortic valve (AoV) and the partially formed interventricular segment of the membranous septum (MS). Because of the failure of formation of the right ventricular inflow tract in the specimen shown in (B), the interventricular segment of the membranous septum is lacking. The foramen is bounded instead by the infundibular septum (IS). Note also the difference in orientation between the posterior interventricular septum (PS) and the bulboventricular septum (BVS). Other abbreviations as above.
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A specimen with right atrial orificial atresia and left-sided outlet chamber the left atrioventricular valve was morphologically a mitral valve (Fig. 12).

C. VENTRICULOARTERIAL CONNECTIONS
All the 6 hearts with atroventricular concordance (right atrium separated from right ventricle by imperforate membrane) had normal ventriculoarterial connections, albeit with pulmonary atresia in 4 cases. Connections were very variable when the ventricular morphology was that of primitive ventricle (Fig. 4). Normal connections (60 of 77 hearts) were most frequently encountered, usually with normal relations (58 of 60). Four of these hearts had pulmonary atresia. Two of the hearts with normal connections had l-malpositioned aortae (anatomically corrected malposition). Twelve hearts had transposition, 10 with d-transposed aortae. It was noteworthy that 1 of 2 cases with l-transposition had a left-sided mitral valve. The 3 hearts without outlet chambers had double outlet chambers by definition, 2 with d- and 1 with l-malposition. One additional heart had a double-outlet connection, from the outlet chamber. One specimen had a persistent truncus arteriosus.

D. PULMONARY AND AORTIC STENOSIS
The most frequent source of stenosis was a constrictive bulboventricular foramen in hearts with primitive ventricle. Such a constriction produced

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**Fig. 12** Photographs of a heart with primitive ventricle outlet chamber, right atrial orificial atresia, and l-transposition. (A) Anterior view of the heart. The outlet chamber is anterior to the main chamber and the aorta is left sided. The covered outlet chamber is seen in (B). It is small and the bulboventricular foramen opens directly beneath the aortic valve. (C) The blind-ending right atrium. Note the prominent venous valve (RVV). (D) Main chamber opened from behind and from the left. Note the typical mitral valve and the prominent posterior ridge (PR). (E) Viewed from the right through the cut visible in (D). The front of the mitral valve is seen with pulmonary-mitral valvar continuity. Despite the presence of right atrial orificial atresia and a left-sided mitral valve the aorta and bulboventricular foramen (BVF) are both left sided. Other abbreviations as above.
pulmonary stenosis in 30 hearts with normally connected arteries, and aortic stenosis with associated isthmal hypoplasia in 5 hearts with transposition (Fig. 14). Another source of pulmonary stenosis in the presence of normal connections was a constriction between the trabeculated and infundibular segments of the outlet chamber; this was seen in 4 hearts in addition to stenosis at the bulboventricular foramen. Valvar stenosis also frequently accompanied bulboventricular stenosis but in 3 cases was the sole cause. In one further case, fibrous tissue from the underside of the infundibular septum obstructed the bulboventricular foramen. Pulmonary stenosis was not seen in association with transposition.

E. ADDITIONAL ANOMALIES
Juxtaposition of the auricles was encountered in 4 hearts. Three had 1-malpositioned aortae, 2 of these with normal ventriculoarterial connections (anatomically corrected malposition).

Discussion
Our investigation has shown, as others have found before (Okin et al., 1969; Van Praagh et al., 1971;
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Fig. 14  Diagram showing that the physiological consequences of bulboventricular construction depend upon ventriculocardinal connections. When the arteries are normally connected, pulmonary stenosis will be produced. In the presence of transposition, aortic stenosis will ensue with the likely sequel of aortic isthmal hypoplasia.

Rao et al., 1973) that there is considerable variation in ventricular morphology in hearts with atresia of the right atrioventricular orifice. Furthermore, as shown by both Henry and Danielson (1974) and Quero-Jimenez et al. (1975) there are further variations in ventricuolarterial connections than merely 'normal relations' or 'transposition'. The question is then posed as to how all these variations can best be described. We believe that adequate description and classification can only be produced by use of the segmental approach (Van Praagh, 1972; Shirebourne et al., 1976). However, it became evident that application of such a segmental approach gave rise to many potential controversies.

A. Why Right Atrioventricular Orificial Atresia Rather Than Tricuspid Atresia?
In this investigation we have described hearts in which the right atrial chamber was blind ending. In all but 2 of these hearts the valve present between the left atrium and the ventricular chamber was a morphologically mitral valve. However, in the 2 remaining hearts the left-sided valve bore more resemblance to a morphologically tricuspid valve. The morphological inference to be drawn from this information is that d-bulboventricular looping occurred in all but the 2 hearts with left-sided tricuspid valves, in which l-looping is presumed to have occurred. Would it not therefore be more accurate to separate these 2 hearts as mitral atresia? Morphologically this would be completely acceptable. However, we submit that in clinical terms the important feature is that the right atrium is

blind ending, irrespective of whether the atretic valve is morphologically mitral or tricuspid. Furthermore, we believe that it would not be possible angiographically to distinguish a mitral valve from a tricuspid valve. The clinical evidence for l-looping would be inferential, being deduced from presence of a left-sided outlet chamber. Yet one of the hearts previously studied (Fig. 12) possessed a left-sided outlet chamber supporting an l-transposed aorta; these observations suggest l-looping but the left atrioventricular valve was morphologically a mitral valve. We, therefore, consider it more accurate and less confusing to refer to 'right atrioventricular orificial atresia' (or 'right atrial orificial atresia'). We have thus excluded from this series hearts with evidence of l-bulboventricular looping and left atrial orificial atresia. In morphological terms this condition represents 'tricuspid atresia' (Harris and Farber, 1939; Keith et al., 1958), but we prefer to consider it as left atrioventricular orificial atresia, and we would group it together with other forms of primitive ventricle which exhibit blind-ending left atria (Quero, 1970; Macartney et al., 1976).

B. Can Right Atrioventricular Atresia Coexist with Primitive Ventricle?
As Macartney and his colleagues (1976) have recently indicated, it has become customary to exclude by definition from the category of primitive (single or common) ventricle, cases of atrioventricular valve atresia. They concluded that in terms of angiography this exclusion was unjustified, a view previously put forward by Quero (1970). Our investigation fully endorses their opinions, and shows that the majority but not all of our examples of right atrioventricular valve atresia are more akin to primitive ventricle than to hearts with normal right and left ventricles. Van Praagh et al. (1964) did not expand on their reasons for excluding cases with atrioventricular valve atresia from single ventricle. Lev and his colleagues (1969) believed that the differentiation depended on whether the main chamber contained the undeveloped right ventricular inflow portion in a position in which it was related to the atretic orifice. They conceded the possibility that this might indeed be the case in hearts with transposition, as previously suggested by Edwards et al. (1965). However, they stated that this was not the case with normally connected arteries, citing as evidence the fact that conducting tissue studies had demonstrated a right bundle-branch entering the outlet chamber. Our own histological observations are in agreement with theirs, but our interpretation differs. We have observed a
similar distribution of conducting tissues in the outlet chamber in primitive (single) ventricle (Anderson et al., 1974a). Furthermore, our histological studies in right atrioventricular orificial atresia have shown that the atretic orifice does indeed 'point' to the main chamber. This fact indicates to us that the hearts thus fulfil the criterion demanded by Lev et al. for single ventricle, since we believe that the right ventricular inflow portion is indeed contained within the main chamber, which therefore constitutes a primitive ventricle. As in 'single' or 'common' ventricle with outlet chamber, the outlet chamber represents the embryonic bulbus (Keith, 1906). We suggest that in primitive ventricle with two atroventricular valves, the right inflow portion has developed with persistence of the bulboventricular septum in its initial anterior position, whereas in right atrioventricular orificial atresia there has been lack of development of the inflow portion but persistence of the anteriorly situated bulboventricular septum (Fig. 15). This concept is supported by the previous study of Rosenquist et al. (1970), who showed most elegantly that the atretic right atrioventricular orifice was potentially in communication with the 'left' ventricle rather than the 'right'. In view of this evidence, it seems to us proper that the definition of primitive (single or common) ventricle should be modified as suggested by Macartney et al. (1976). Thus, if primitive ventricle exists when both atria are either actually or potentially in communication with the same ventricular chamber, then the majority of specimens of right atrioventricular orificial valve atresia can properly be classified as primitive ventricle (Anderson et al., 1976).

It must be emphasised once more that not all cases of right atrioventricular orificial atresia are cases of primitive ventricle. Six of our cases had true right ventricles. Four of these had right ventricular hypoplasia, comparable with the cases reported by Okin et al. (1969). The remaining cases had imperforate Ebstein's anomaly of the right atrioventricular valve. This rare condition was well documented by Rao et al. (1973) who reviewed previous cases and emphasised the clinical importance of its recognition.

C. ASSOCIATED ANOMALIES

The clinical importance of the classification of Edwards and Burchell (1949) was that it described the state of pulmonary blood flow. In order to convey this important information, Henry and Danielson (1974) tabulated their extensive findings, using a modification of this approach. However, the segmental approach does not preclude the provision of this information if it is appreciated that pulmonary stenosis is an associated anomaly. An important and vital part of segmental nomenclature is the tabulation of additional anomalies. In this way other information will also be transmitted and attention focused on the genesis of these associated anomalies. Thus pulmonary stenosis with right atrioventricular orificial atresia is most frequently the result of a constrictive bulboventricular foramen. If one thinks segmentally it is readily apparent that a similar constriction in the presence of transposition will result in aortic stenosis and associated isthmal hypoplasia. When pulmonary stenosis occurs with transposition, it must have a different cause, usually posterior deviation of the infundibular septum.

D. SURGICAL SIGNIFICANCE

With the demonstration by Fontan and Baudet (1971) that radical palliative surgery is possible in the presence of right atrial orificial atresia, knowledge of all aspects of anatomy is vital. It will immediately be evident that hearts with formed right ventricles do not require a Fontan-type procedure. In one of the
Right AV valve atresia

hearts we have described, excision of an imperforate Ebstein anomaly would have restored the normal circular pattern (Fig. 6). Rao et al. (1973) have previously commented on the surgical importance of recognizing this variety of right atrial orificial atresia. It is because of the surgical implications that we have excluded from this series hearts with left atrial orificial atresia even when the atric valve was morphologically tricuspid. Since the Fontan operation can be employed only when the right atrium is blind ending, we prefer to distinguish between right and left atrial orificial atresia, rather than between tricuspid and mitral atresia.

The fact that a small number of the hearts were examples of primitive ventricle without outlet chamber may also be significant. It is becoming customary now to insert the distal end of the right atrial conduit to the outlet chamber. Clearly this would be impossible when an outlet chamber is not present. In the hearts we studied an outlet chamber was also effectively lacking when there was pulmonary atresia with normal ventriculoarterial connections. However, in all other hearts a good size outlet chamber was present even when secondary constriction of the bulboventricular foramen had occurred. In 4 hearts a second constriction was present distally within the outlet chamber which may have detracted from its functional value: as a potential pumping chamber if the distal end of the conduit had been inserted to that chamber.

The type of ventriculoarterial connection present also influences the site of insertion of the distal end of the conduit. The outlet chamber can only be employed as part of the pulmonary circuit in the presence of normally connected arteries. It is important to realise, therefore, that normal ventriculoarterial connections can exist with an anterior left-sided aorta. This relation is usually termed anatomically corrected malposition, and its association with right atrial orificial atresia has previously been stressed by Melhuish and Van Praagh (1968) and by Freedom and Harrington (1974). It must also be remembered that any possible ventriculoarterial connection can exist with right atrial orificial atresia.

Conclusions

In summary, therefore, we believe that our study has shown a basic subdivision in terms of ventricular morphology in hearts with atresia of the right atroventricular orifice, the majority being examples of primitive ventricle with outlet chamber. We believe that understanding of the condition is enhanced if the anomaly is described as right atrial orificial atresia rather than ‘tricuspid atresia’ since in some hearts the atretic orifice may be a mitral orifice, but nevertheless its effect is to shunt systemic venous return to the left atrium. Finally, we believe that our investigation endorses once more the value of a segmental approach to diagnosis (Van Praagh, 1972; Shinebourne et al., 1976), since many variants of right atrial orificial atresia are described, while others probably exist which have yet to be described.

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References


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