Coexistence of Single Ventricle with Atresia of One Atrioventricular Orifice

By Manuel Quero

SUMMARY
A new case of an unusual association of mitral atresia with single left ventricle is reported. The salient anatomic features include: (1) the right atrioventricular valve is atretic; (2) the left atrioventricular valve enters the large ventricle with morphologic characteristics of the left ventricle; and (3) this large ventricle communicates by way of an interventricular communication with a smaller left-sided ventricle, but with the morphologic characteristics of the right ventricle. Guidelines for the recognition of these uncommon malformations are given.

Additional Indexing Words:
- Single left ventricle
- Double-inlet left ventricle
- Atrioventricular orifice atresia
- Mitral atresia
- L-Loop

The single left ventricle\(^1\) or single (primitive) ventricle\(^2\) has been defined as an anomaly of the heart in which both atrioventricular orifices, either separated or fused into a common atrioventricular canal, are received by a left ventricle. The right ventricular cavity, with a more or less pronounced hypoplasia of its sinusus portion, is reduced in some cases to its outflow tract.

The term double-inlet left ventricle proposed by Mehrizi et al.\(^3\) and de la Cruz and Miller\(^4\) defines cases of similar malformations with separate atrioventricular orifices. The latter postulate by their embryologic hypothesis that this malformation could result from an arrest in the normal movement of the atrioventricular canal toward the bulbus cordis.

Cases with atresia of one atrioventricular orifice have been excluded from the above-mentioned malformations.

To avoid equivocal reports of the coexistence of the double-inlet left ventricle with atrioventricular orifice atresia, the concept of a "topographically homologous ventricle" was introduced in a recent report.\(^5\) (Note: In a heart with atresia of either atrioventricular valve, in which the possible existence of a single left ventricle has to be proved or ruled out, we consider the topographically homologous ventricle to be the one that would be expected to be situated beneath the atretic valve.) A recent contribution\(^6\) dealing with these malformations includes cases with mitral atresia without detailed descriptions of the specimens.

The following case makes a new observation of this uncommon association.

Case Report
A 3-month-old female patient expired from cyanosis and congestive heart failure. Chest roentgenograms showed severe cardiac enlargement and increased pulmonary vasculature, and the electrocardiogram revealed right atrial hypertrophy and a stereotyped rS pattern over the entire precordium.

Autopsy Observations
The right atrium receiving the two venae cavae
Single Ventricle with Atresia

Internal view of the right atrium. Note the atretic (*) atrioventricular orifice and the patent foramen ovale (FO).

was greatly dilated and hypertrophied. The left atrium and its tributary veins, the pulmonary veins, were fairly dilated. A right atrioventricular orifice atresia prevented direct communication between the right atrium and the ventricles (fig. 1). The blood flowing from the two venae cavae was forced through a foramen ovale and the left atrium and a greatly dilated ventricular cavity with morphologic characteristics of a left ventricle (fig. 2). The situation of the left ventricle to the right of the smooth septal surface and the existence of the infundibular chamber to the left of this septum brought about the conclusion that the overall disposition of the ventricular cavities be considered as of the L-loop type. The elongated infundibular chamber (fig. 3), almost horizontally inclined toward the right, supported an anterior right-sided aorta. The bulboventricular inversion coexisting with the anterior right-sided aorta was thought to be due to the grossly abnormal horizontal inclination of the infundibular chamber (fig. 3).

Discussion

In the usual case of mitral or tricuspid atresia the main anatomic lesion, the atresia, would conform with the hypoplasia or aplasia of the "topographically homologous ventricle."5 Left and right atrioventricular orifice atresias in cases of L-loop are associated with hypoplastic topographically homologous ventricles with reversed morphologic characteristics.5

Below, two cases of atrioventricular orifice atresia with the topographically homologous ventricle, enlarged instead of hypoplastic (the hypoplastic chamber being the other ventricular chamber), are dealt with.

One of these two cases (fig. 4E), previously described by the author,5 sets the premise for the discussion of the second case, which is the object of this paper (fig. 4F).

Figure 4E represents a heart which has a D-loop pattern, with a right-sided ventricular cavity possessing morphologic characteristics of a right ventricle and a left-sided ventricular chamber with anatomic features of a left ventricle. It is surprising that, despite the existence of a left atrioventricular orifice atresia, the left-sided ventricular chamber is enlarged instead of being hypoplastic and the right ventricle is reduced to its outflow tract–ventricular proportions which are characteristically found in cases of single left ventricle or double-inlet left ventricle. The
Internal view of the right-sided left ventricular cavity (LV). Note the smooth appearance of the upper part of the left septal surface (VS) and the delicate trabeculation of its lower part. The left atrioventricular orifice (LAVO), in this case of an L-loop, the tricuspid orifice, is entirely received by the left ventricle (LV). The leaflets of the left atrioventricular valve (avv) are supported by three papillary muscles. Two of them (pm) are situated within the left ventricle (LV) and the third (pm') is attached to the top border of the ventricular septum (VS) partially obstructing the communication (F) between the left ventricle and the infundibular chamber. The pulmonary artery (PA), posteriorly situated in relation to the aorta (Ao), originates directly from the left ventricle (LV), its valves remaining in fibrous continuity (*) with the atrioventricular valves (avv). The aorta (Ao), anteriorly situated, originates at the left side of the conal septum (CS) and the ventricular septum (VS) from a left-sided infundibular chamber. raa = right atrial appendage.

Figure 2

reason for clarifying these apparent discordant findings is that the right atrioventricular orifice instead of opening into the right ventricle is received by the left ventricular chamber, possibly because during the embryo development it was not moved from the primitive ventricle to the bulbus cordis, and it remained connected with the left ventricle as it seems to occur in single left ventricles or double-inlet left ventricles.\(^4\) Bearing in mind the foregoing, the ventricular proportions of this case should occur despite the presence of a left atrioventricular orifice atresia.

In the second case, an atretic right atrioventricular orifice was associated with an enlarged left ventricle and an infundibular chamber (fig. 4F, left). The aorta was right sided and anterior to the pulmonary artery (fig. 4F, right).

Apparently with this description nothing of interest would be present in this case, but a closer examination revealed that, although an anterior and right-sided aorta existed, the bulboventricular loop was an L type, the infundibular chamber being situated to the left of the main left ventricular chamber. The discordance between the situation of the aorta and the type of bulboventricular loop is attributed to the existence of an elongated infundibular chamber almost horizontally inclined toward the right.

By reason of the presence of an L bulboventricular loop the correct interpretation of this case is made to vary considerably. Far from being a simple case of tricuspid atresia with transposition of the great arteries, it is considered to be a bulboventricular inversion with a right atrioventricular (mitral) atresia. In this case, where one would have expected the left ventricle to be hypoplastic, it is rather surprising that this cavity appears greatly enlarged and the right ventricle, the one expected to be enlarged, is reduced to an infundibular chamber (fig. 4F). The reason for explaining this apparent discordance is that, as in the first case, the tricuspid orifice has not moved to the bulbus cordis, remaining mainly connected with the left ventricle (fig. 4F, left).

One difference between our two cases is that the movement of the tricuspid orifice toward the bulbus cordis, absent in case 1, was initiated to a slight degree in case 2. Stemming from this, the infundibular chamber in the first case neither sheltered papillary

Circulation, Volume XLVI, October 1972
Internal view of the right ventricular remnant or infundibular chamber. Note how the aortic valves (AoV) are separated from the atrioventricular valves (AVV) by a muscular band, the conal septum (CS). A papillary muscle (p'm) attached to the top of the ventricular septum gives some support to the atrioventricular valve (AVV). Ao = aorta; CO = coronary ostia. Notice the horizontal inclination of the infundibular chamber.

DISCORDANT SITUATIONS—WITH AN ENLARGED TOPOGRAPHICALLY HOMOLOGOUS VENTRICLE, ABSENCE OF THE NORMAL DISPLACEMENT OF THE ATRIO-VENTRICULAR CANAL TOWARDS THE BULBUS CORDIS.

D-LOOP—MITRAL ATRESIA AND LACK OF THE NORMAL DISPLACEMENT OF THE “TRICUSPID PORTION” OF THE ATRIOVENTRICULAR CANAL TOWARDS THE BULBUS CORDIS. Despite the L-LOOP, the infundibular chamber, almost horizontally directed towards the right, supports an anterior and right-sided aorta.

Figure 3

Illustrations showing the two cases observed by the author in which a mitral atresia coexisted with a big left ventricle and an infundibular chamber (a single left ventricle or double-inlet left ventricle). See text. SVC = superior vena cava; IVC = inferior vena cava; RA = right atrium; PV = pulmonary veins; LA = left atrium; RRV = remnant of the right ventricle; IC = infundibular chamber; LV = left ventricle; L.A.V.O. = left atrioventricular orifice; R.A.V.O. = right atrioventricular orifice; T.O. = tricuspid orifice; “RV” = left-sided right ventricle; “LV” = right-sided left ventricle; R”RV” = remnant of a left-sided right ventricle.

Figure 4
muscles not chordae, both of these structures being present in the second case.

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Circulation. 1972;46:794-798
doi: 10.1161/01.CIR.46.4.794

Circulation is published by the American Heart Association, 7272 Greenville Avenue, Dallas, TX 75231
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Print ISSN: 0009-7322. Online ISSN: 1524-4539

The online version of this article, along with updated information and services, is located on the World Wide Web at:
http://circ.ahajournals.org/content/46/4/794

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