Atresia of the Left Atrioventricular Orifice
Associated with a Holmes Heart

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SUMMARY
The first well-documented case of Holmes heart associated with an atretic left atrioventricular orifice is reported. The Holmes heart may be considered as a single ventricle in which the main chamber has the morphologic characteristics of a left ventricle, the infundibular chamber is not inverted, and the great arteries are normally arranged. The importance of this report is greater than its simple description since it definitely establishes that coexistence of a true single ventricle and atresia of the atrioventricular orifice is possible.

Additional Indexing Words:
Single ventricle Common ventricle Double-inlet left ventricle
Hypoplastic left ventricle Infundibular chamber Mitral atresia
Primitive ventricle

IN THE MAJORITY of cases of the isolated form of mitral atresia the left ventricular cavity is hypoplastic or even aplastic and the right ventricle is huge. In other cases, the left ventricle is not hypoplastic, and examples of normal or even enlarged left ventricular cavities have been described. A greatly enlarged right ventricle is, however, the rule. Examples of mitral atresia with even minor degrees of hypoplasia of the right ventricle have not been reported.

The single ventricle, type A of Van Praagh and associates, corresponds to the single (primitive) ventricle of Lev's group. Both designations can be given interchangeably to a rather distinct pathological entity, characterized by a morphologically left ventricular chamber which receives the blood from the two atria, either through separated valves or a common atrioventricular valve. One of the two great arteries emerges directly from this ventricle, between the two atrioventricular valves (or between the two components of the common atrioventricular valve) in total or partial fibrous continuity with one or both atrioventricular valves. This great vessel is the aorta in cases without transposition and the pulmonary artery when the great arteries are transposed. The other great artery takes its origin through a ventricular septal defect (more properly called the "bulboventricular foramen") and an infundibular chamber to the right or to the left of the main ventricular chamber, according to the type of the bulboventricular loop.

In this type of single ventricle, the right ventricular sinus is considered to be absent. At least, it must not be present in the infundibular chamber. Therefore, no papillary muscles, chordae, or well-recognizable posterior portion of the ventricular septum must be present in the infundibular chamber.

Recently, de la Cruz and Miller and Mehrizi and co-workers have described a rather similar pathologic entity, the double-inlet left ventricle. The first two authors presented two pathologic specimens in which the morphologically left ventricle contained the two atrioventricular ostia. In their first

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case, the tricuspid orifice overrode the ventricular septum, and its anterior leaflet was inserted in the remnant of the right ventricle, which contained papillary muscles. In their second case no papillary muscles were seen in the right ventricle; nevertheless, a small portion of the ventricular septum could be recognized.

In the strictest sense, therefore, neither of these cases can be included under the designations given by Van Praagh and Lev and their co-workers, since in them, some traces of the existence of a right ventricular sinus were present. Nevertheless, it is not difficult to conceive the possible existence of cases of double-inlet left ventricle with the right atrioventricular orifice completely contained in the left ventricular chamber and, possibly, with complete aplasia of the right ventricular sinus. These cases, of course, will be identical with those included under the designations of Van Praagh and Lev given before. In such cases, the question of whether we use one designation or another is an unimportant semantic problem. The Holmes heart is a single ventricle, type A of Van Praagh or a single (primitive) ventricle of Lev in which the infundibular chamber is not inverted and the great arteries are normally arranged.

To the present time, all the pathologic situations described above, have been reported without being associated with atresia of an atrioventricular orifice. Moreover, atrioventricular orifice atresias have been excluded in the definitions of these anatomic complexes. The reason may be that a case like the one which is the object of this report has not been previously observed. For this reason, we think, our case deserves to be published.
ATRESIA OF LEFT A-V ORIFICE

Figure 3
Anterior view of the heart showing moderate cardiac enlargement and normally related great arteries. A = aorta; P = pulmonary artery.

Figure 4
Interior view of the left atrium illustrating the atresia of the mitral orifice (asterisk) and the probe-patent foramen ovale (FO).

Report of Case

The patient, a 3-month-old female, was hospitalized because of cyanosis since birth and anoxic spells. On examination, she was deeply cyanosed, with tachypneic respirations. Peripheral pulses were all palpable. Heart rate was 170/min. The liver edge was at a normal level. Palpation of the precordium disclosed a moderately enlarged heart. A faint systolic ejection murmur and a single second sound were heard.

Chest roentgenograms (fig. 1) revealed slight cardiac enlargement and pulmonary venous congestion. The gastric bubble and liver shadow were normally situated. The electrocardiogram (fig. 2) showed a mean QRS axis of +70°. A QS complex was recorded in lead aVn, and rS deflections were visible over the entire precordium. Conspicuous ST-T changes were noted in the left precordial and frontal plane leads.

A clinical diagnosis of serious hypoxemia, probably due to a severe and complicated form of hypoplastic left heart.
congenital heart disease, was made, and conservative medical treatment with oxygen and tromethamine with electrolytes (Tham-E) was instituted. Arrangements for cardiac catheterization and angiography were going on, when the patient suddenly died with an irreversible cardiac and respiratory arrest.

Autopsy Observations

The heart was moderately enlarged. Externally (fig. 3) the two atria and their tributary veins seemed to be normal, both topographically and morphologically. The great arteries were normally related (fig. 3). The right coronary artery emerged from the right sinus of Valsalva. It gave off the right infundibular delimiting branch and the right circumflex. The left coronary artery emerged from the left sinus of Valsalva and gave off the left delimiting coronary artery, a descending branch, similar to the usual anterior descending branch, and the left circumflex. A posterior descending branch was not visible.

The left atrium was moderately enlarged. There was no mitral orifice, the only channel for the flow of the pulmonary venous blood being a probe-patent foramen ovale (fig. 4). The right atrium opened into a ventricular chamber which had the morphologic characteristics of a left ventricle (fig. 5). The atrioventricular valve between these two cavities was a bicuspid valve; two groups of papillary muscles—anteriord and posterior—supported the two leaflets of this valve.

The aorta took off from this ventricle and was only in partial fibrous continuity with the single atrioventricular valve (fig. 5). High in the ventricular septum, a bulboventricular foramen (fig. 5) led into a very narrow, non-inverted infundibular chamber from which the pulmonary artery arose (fig. 6). In this infundibular chamber, no papillary muscles, chordae, or a well-recognizable posterior portion of the ventricular septum were present (fig. 6).

Discussion

Since the original definition by Keith, some of the most remarkable descriptions about single or common ventricles have not included cases of atresia of the atrioventricular orifices. Nevertheless, the same series includes cases of stenosis of either atrioventricular valve and, in one of the last studies, a case of single ventricle with a severely stenotic atrioventricular valve, finally becoming atretic, is reported.

On the other hand, some very valuable reviews on the subject include, among the cases single or common ventricles, cases of associated atresia of either atrioventricular orifice. In these last studies however, little attention has been paid to the morphologic characteristics of the ventricles and the type of the bulboventricular loop. Consequently, a critical exclusion of other possibilities, such as an extreme hypoplasia or aplasia of the "topographically homologous ventricle" has been lacking.

In a heart with atresia of either atrioventricular valve, in which the possible existence of a single 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.

For example, in a case of atresia of the left atrioventricular orifice without inverted infundibulum, the topographically homologous ventricle would be the morphologically left ventricle. Conversely, in a specimen with a left atrioventricular orifice atresia and an inverted infundibulum, the topographically
homologous ventricle would be the morphologically right ventricle.

Similarly, in cases of right atrioventricular orifice atresias, the topographically homologous ventricle would be the morphologically right ventricle in hearts without an inverted infundibulum and the morphologically left ventricle in specimens with an inverted infundibular chamber.

In this way, cases in which the atrioventricular orifice is atretic and the topographically homologous ventricle is small, but recognizable, are not necessarily examples of true single ventricles, from an anatomic point of view. They can simply be cases of ventricular hypoplasia. Such is the situation, for example, in the usual case of mitral atresia with normal aortic root or in the more complicated cases of mitral atresia with a double-outlet right ventricle.

In the same way, cases with entirely aplastic “topographically homologous ventricles” must not be necessarily considered as single ventricles, although they cannot be definitely ruled out as such. For example, cases of left atrioventricular orifice atresia, without inverted infundibula, with grossly enlarged right ventricles from which the two great arteries take their origin, and without recognizable left ventricular chambers, are not necessarily true single ventricles. They can be true single ventricles, but until better means of recognizing the ventricles or their remnants are available, it is better to wait for their exact anatomic diagnosis.

However, cases in which either atrioventricular valve is atretic and the topographically homologous ventricle is completely present and, the other ventricle is absent or incomplete, are very probably cases of true single ventricle, in which atresia of the atrioventricular orifice coexists.

The topographically homologous ventricle in our case—a case of left atrioventricular orifice atresia without infundibular inversion—would be the morphologically left ventricle. Is it present? One can state that it is the only one entirely present; the morphologically right ventricle exists only in its infundibular cham-

ber. If an inverted septum and infundibulum had been present, the case would have been classified as a corrected transposition with tricuspid atresia. In this situation, the topographically homologous ventricle would have been the morphologically right ventricle, and the one that is incompletely developed would have been the hypoplastic one.

To make sure, then, that a supposed single ventricle with atresia of the atrioventricular orifice is necessarily a true single ventricle from an anatomic point of view, it is convenient to analyze its morphology and the type of bulboventricular loop that each particular example displays. If these important features are overlooked, it is not possible to state, with certainty, that such specimens are true single ventricles. In this respect, to my knowledge, this process has been done for the first time in this case. Consequently, I think this case is the first reported example in which there can be no argument against the hypothesis, that this is indeed a case of single ventricle (or a double-inlet left ventricle) with atresia of the left atrioventricular orifice.

There is not enough evidence to assure the existence of a right ventricular sinus since the remnant of the right ventricle does not contain papillary muscles, chordae, or a well-recognizable posterior portion of the interventricular septum. We consider this case, therefore, an example of a single ventricle (type Al of Van Praagh or single, primitive ventricle of Lev) rather than a double-inlet left ventricle, since in the two cases reported by de la Cruz and Miller, some elements were found that indicated the presence of a right ventricular sinus. Nevertheless, as stated before, these two pathologic entities can be so similar, that no attempt will be made to rule out the possibility of this case being a double-inlet left ventricle with a left atrioventricular orifice atresia.

We have deliberately used the term "left atrioventricular orifice atresia" instead of mitral atresia, since in these situations, the two atrioventricular valves are often bicuspid and, therefore, the terms "mitral" and "tricuspid" become meaningless.
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