Morphologic Spectrum of Ventriculoarterial Connection in Hearts With Double Inlet Left Ventricle: Implications for Surgical Procedures

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Background. **This study was conducted to determine a morphologic spectrum of ventriculoarterial connection in double inlet left ventricle and implications for surgical procedures.**

Methods. **Examined were 54 autopsied heart specimens and 43 consecutive clinical patients.**

Results. **The hypoplastic and incomplete morphologic right ventricle was located leftward to the dominant ventricle in 62 and adjacent to the right atrium in 35. Common patterns were seen in 46 of 62 (74%) with the right ventricle leftward (discordant ventriculoarterial connection with the aorta left-anteriorly located ["SLL" type]) and in 28 of 35 (80%) with the right ventricle rightward (either the normally connected great arteries in 13 or discordant connections with the aorta right-anteriorly located in 15). In the remaining 23 hearts, the great arteries were unusually oriented in 7, the outlet septum was malaligned in 9, or the pulmonary trunk was atretic in 7. In those with malalignment, the ventriculoarterial connections were double outlet from the right ventricle, from the left ventricle, or were transitional with overriding of one of the great arteries. In the clinical series, 19 of 35 patients (54%) in whom the aorta arose from the morphologically right ventricle underwent either myectomy to enlarge the interventricular communication or a Damus-Kaye-Stansel anastomosis was fashioned to treat existing or potential subaortic stenosis. Only 1 of 8 patients with the aorta arising from the dominant ventricle needed similar surgical procedures.**

Conclusions. **Ventriculoarterial connection in double inlet left ventricle demonstrated a morphologic spectrum and needs precise recognition to provide an unobstructed ventricular outflow after operation.**

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In the setting of double inlet left ventricle, discordant ventriculoarterial connection is commonly seen in hearts with a hypoplastic and incomplete morphologically right ventricle on the left side (the so-called SLL type) [1–3], whereas a concordant ventriculoarterial connection, which is a vital element for the so-called Holmes heart, is less common [4, 5]. A small proportion of hearts with double inlet left ventricle have some other manner of ventriculoarterial connections, such as double outlet from the right or the left ventricle [6, 7]. Accordingly, we investigated a combined series of hearts with this entity, in postmortem specimens and in patients, focusing on ventriculoarterial connections to determine precisely the morphologic spectrum and to consider implications for options available for surgical procedures.

Material and Methods

This retrospective study was approved by the Institutional Ethics Committee, and individual patient consents were waived.

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We examined 54 autopsied heart specimens of double inlet left ventricle. Another consecutive series of 43 clinical patients were assessed on the basis of imaging as well as direct inspection during heart operations. All patients underwent an intracardiac operation, and none were included in the autopsied series. The type of definitive repair was ventricular septation in 22 and the Fontan type procedure in 19. One patient remained with the bidirectional Glenn physiology, and another patient with coarctation of the aorta underwent the Norwood type procedure.

In total, 96 had the usual atrial arrangement, and only 1 clinical patient possessed its mirror imagery. This exceptional case was described as seen in a mirror so that we could correlate it with the other cases. The incomplete and hypoplastic morphologically right ventricle was located anteriorly and to the left of the dominant left ventricle in 62, but rightward and adjacent to the right atrium in 35.

Results

The morphologic features around the ventriculoarterial connection are summarized in Table 1.

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Table 1. Summary of Morphologic Features Around the Ventriculoarterial Connections

Feature	RV Leftwards	RV Rightwards
Ventriculoarterial connection[a]		
Concordant	2	13
Discordant	51	16
Double outlet RV	1	3
Aorta from RV/pulmonary atresia	6	1
Double outlet LV	2	1
Aorta from LV/pulmonary atresia	. . .	1
Infundibular morphology		
Subpulmonary	2	13
Subaortic	58	17
Subpulmonary & subaortic	1	3
Markedly attenuated	1	2
Aortic valve in relation to pulmonary valve		
Right posterior	2	14
Right anterior	2	17
Right side-by-side	2	3
Left anterior	54	. . .
Left side-by-side	2	1
Arterial trunks		
Spiraling	2	14
Parallel	60	21

[a] For overriding of the aortic or the pulmonary valve, the so-called 50% rule was applied.

LV = morphologic left ventricle; RV = morphologic right ventricle.

Common Types

Of 62 with the right ventricle leftwards, 46 (74%) had the SLL type, in which the left-anteriorly located ascending aorta originated entirely from the morphologically right ventricle with the pulmonary trunk (right posterior) entirely from the dominant left ventricle (Fig 1). Of 35 with the right ventricle rightwards, 13 (37%) had the Holmes heart, with the normally related great arteries with concordant ventriculoarterial connection, and 15 (43%) had discordant ventriculoarterial connection with the aorta located right anteriorly to the pulmonary trunk (Fig 2).

Atypical Patterns in Hearts With the Right Ventricle Leftwards

Four (6%) had discordant ventriculoarterial connections but with unusual aortopulmonary orientations (Fig 1); the aorta was to the left of the pulmonary trunk in a side-by-side fashion in 2 or was right-anteriorly located in 2. The outlet septum in these 4 hearts was in alignment with the muscular ventricular septum; in another 4, the outlet septum was malaligned (posteriorly oriented in relation to the muscular ventricular septum). Because of that, the aorta was overriding the left ventricle by 25% in 2, by 50% in 1, or exclusively arising from the left ventricle in the remaining 1. In these hearts the aorta was anterior or left anterior to the pulmonary trunk. No heart had a concordant ventriculoarterial connection when the aorta was located anteriorly to the pulmonary trunk; this particular situation is known as a part of the features that are seen in the so-called anatomically corrected malposition.

In one heart in which the aorta was located to the right and side-by-side to the pulmonary trunk, both great arteries arose entirely from the incomplete and hypoplastic morphologic right ventricle. The outlet septum was unequivocally malaligned with the muscular ventricular septum. In another 2 with the aorta normally orientated relative to the pulmonary trunk, the ventriculoarterial connection was concordant, and there was no malalignment of the outlet septum. These 2 hearts had a regular spiraling arrangement of the great arteries.

In the remaining 6 hearts (10%), there was pulmonary atresia, with the aorta arising entirely from the morphologic right ventricle, including 1 with an imperforate pulmonary valve (Fig 3). The remnant of the pulmonary trunk suggested a left anterior orientation of the aorta in 5 and right side-by-side in 1.

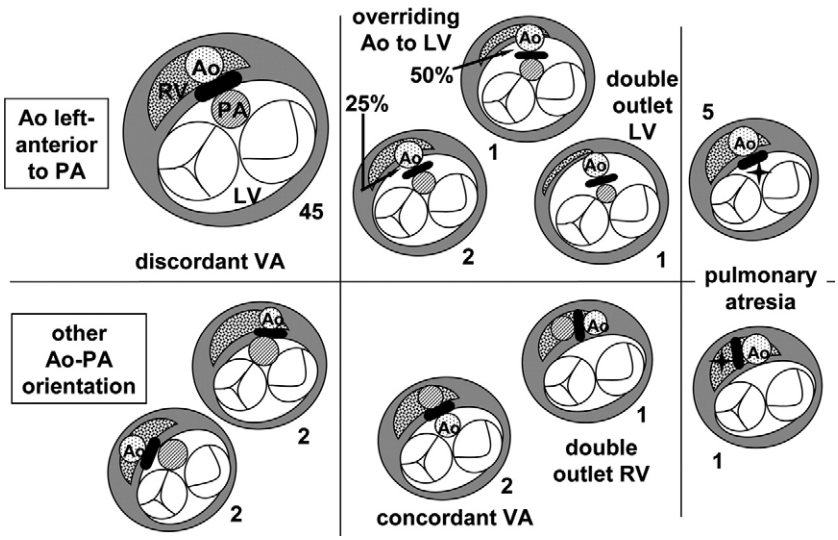

Fig 1. Diagrams of ventriculoarterial connections in patients and specimens with double inlet left ventricle with the right ventricle leftwards in relation to the dominant left ventricle. The schemas are drawn as seen from the top. The numbers of examples are shown adjacent to each schema. Percentages indicate degree of overriding of the aorta. (Ao = aorta; LV = morphologic left ventricle; PA = pulmonary arterial trunk; RV = morphologic right ventricle; VA = ventriculoarterial connection.)

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