Single (Primitive) Ventricle

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SUMMARY

Single ventricle is that condition in which both atrioventricular (A-V) orifices enter a common ventricular sinus, from which the arterial trunks emerge with their coni. The following types of single ventricle were found: (1) with regular (noninverted) transposition in levocardia; (2) with inverted transposition in levocardia; (3) with normal position of arterial trunks in levocardia; (4) in dextrocardia, and (5) in mesocardia. This entity is differentiated from common ventricle which is basically a heart with a large ventricular septal defect. Single ventricle does not include mitral and tricuspid atresia by definition, nor does it include straddling tricuspid orifice, in which two distinct ventricles are present. The concept of single ventricle herein defined differs from de la Cruz and Miller's concept of double-inlet left ventricle which includes straddling tricuspid orifice. Embryologically in single ventricle we are dealing with persistence of the primitive state of the bulboventricular loop, due to a lack of expansion of the atrial canal to the right during the process of absorption of the bulbus.

Additional Indexing Words:

Common ventricle Embryology Double-inlet left ventricle

SINGLE VENTRICLE is defined, in this work, as that condition in which both atrioventricular (A-V) orifices enter, or a common A-V orifice enters, a common ventricular sinus, from which the arterial trunks emerge with their coni. One or both of these coni are sufficiently well developed to comprise a small outlet chamber, or a conus may simply form an outflow tract to the main chamber. On gross examination, this outflow chamber is considered to consist only of the conus of the right ventricle, without any sinus of the right ventricle.

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This paper is an attempt to establish this concept of single ventricle. Toward that end 42 of the 56 hearts seen at the Congenital Heart Disease Research and Training Center which satisfy the criteria just listed for single ventricle were studied anatomically. These were among the 2,745 congenitally abnormal hearts seen in this Center in the last 10 years. To these were added four hearts with single ventricle present in our collection but without autopsy data. The anatomic findings in these hearts with a single ventricle were compared with those of hearts with somewhat similar features, that is, to those with a common ventricle, straddling triscuspid orifice, and mitral and tricuspid atresia.

Definition of Terms

In this work the diagnosis of atria and ventricles is based on the septal morphology.[1] Where the presence of a septum is in question, as in the ventricular mass in single ventricle, in our opinion diagnosis of the identity of a chamber is not justifiable. The *sinus* of a ventricle is that portion which normally contains the tensor apparatus of the A-V valves; in other words, the

papillary muscles and chordae. If such tensor apparatus resembles the normal tricuspid apparatus, it is right sinus, and, if it resembles the mitral apparatus, it is left sinus. If such tensor apparatus is absent, the presence of a sinus may be inferred if the area is demarcated by a well-recognized posterior portion of the ventricular septum. In our opinion there is not sufficient evidence that the trabecular structure of a parietal area provides an adequate basis for identifying the right or left ventricular sinus. The term *common sinus* is used to indicate that the tensor apparatus of both A-V valves is included in that sinus. The term *conus* implies an outflow tract, which, as is known embryologically, is derived from the bulbus. Every arterial trunk, both normally and abnormally, has a conus, however abbreviated or one-sided it may be. The *conal septum* is the muscular septum which forms a separation between the two coni. The position of the coni vis-a-vis the septum may aid in the diagnosis of regular and inverted transposition (see below). The term *small outlet chamber* implies that the conus giving rise to a certain vessel is especially demarcated, and its size approaches that of the normal right ventricular conus. The term *transposition* implies an abnormal relationship either between the arterial trunks from the standpoint of anteroposterior relationships or between the trunks and the atrioventricular orifices. Reference to the origin of the great vessels from specific chambers, used by us ordinarily in transposition, cannot be made in single ventricle since the identity of the chambers is in question. The term *inverted transposition* (Spitzer's[2] term) means that in addition to the disturbance in the anteroposterior relationship of the bulbotruncal area, there is a disturbance in lateralness. The point of reference for judgment of the position of the arterial trunks is the normal position in the trunks seen in levocardia. The terms *right and left delimiting coronary arteries* (Rowlatt's[3] terms) refer to those vessels which demarcate the small outlet chamber. This terminology is necessary in single ventricle, because the term *anterior descending coronary artery* usually cannot be applied to these vessels. To be designated as such, the artery must either lie in the anterior longitudinal groove or supply the anterior two thirds of the ventricular septum. Since there is doubt as to what constitutes a ventricular septum and whether a ventricular septum is present in single ventricle, the term *anterior descending coronary artery* is infrequently used here.

Characteristics and Types of Single Ventricle

The pertinent details of the cases of single ventricle are summarized in table 1.

The classification of the type of single ventricle was based on the direction of the base-apex axis of the heart, the presence or absence of transposition, and the type of transposition. Judgment as to the type of transposition, whether regular or inverted, was made on the basis of the following factors: (1) position of the outlet chamber and its relation to the right and left A-V valves; (2) prominence of the left or right delimiting coronary artery; (3) the relative position of the aorta and pulmonary trunk, and (4) the position of the conal septum vis-a-vis the pulmonary trunk.

All types of single ventricle showed the following characteristics: (1) The mitral and tricuspid valves were not usually readily diagnosable (figs. 1C, 1D, and 2), with one posteromedial and one anterolateral or anterior group of papillary muscles being usually attached to each valve. One of the anterolateral papillary muscle groups tended to be small, or at times absent. (2) The posterior wall of the common sinus was divided by a longitudinal ridge passing from the base of the pulmonary trunk to the apical region (fig. 2). (3) When transposition (regular or inverted) was present and the pulmonary orifice was not atretic, one aspect of the annulus of the pulmonary valve was in continuity with either both A-V orifices (fig. 2) or a common A-V orifice (with one exception).

Single Ventricle with Regular (Noninverted) Transposition in Levocardia (Normal Position of Heart): 15 Cases (Figs. 1 and 2)

In this type of single ventricle, (1) the outlet chamber was situated to the right and was closer to the right A-V valve than to the left; (2) the left delimiting coronary artery was more prominent than the right (with one exception); (3) the left atrial appendage was visible from the anterior view of the heart; (4) the aorta was situated anteriorly and to the right and the pulmonary trunk posteriorly and to the left with two exceptions. In the two exceptional cases, pulmonary stenosis was present, the aorta was anterior and slightly to the left, and the pulmonary trunk was posterior and slightly to the right. Other

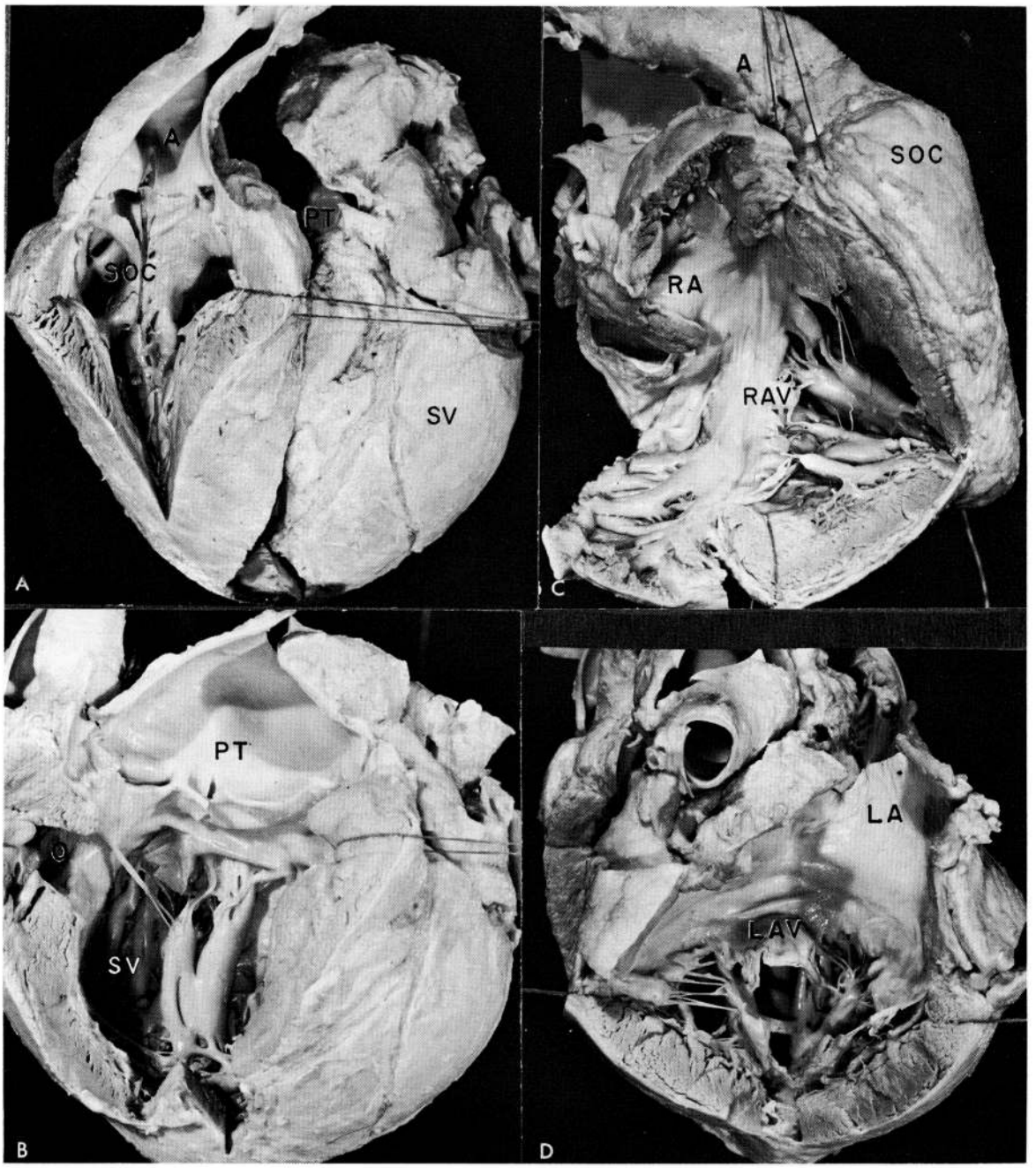

Figure 1

Case 7. Single (primitive) ventricle with regular (noninverted) transposition in levocardia. (A) Anterior view of the heart with the outlet chamber opened. (B) Internal view of single ventricle. (C) View of right atrium and single ventricle. (D) View of left atrium and single ventricle. SOC = small outlet chamber; A = aorta; SV = single ventricle; PT = pulmonary trunk; O = opening into small outlet chamber; RA = right atrium; RAV = right A-V valve; LA = left atrium; LAV = left A-V valve.

characteristics of this type of single ventricle were (5) the atrial septum tended to be more sagittal than it was in the cases of inverted transposition; (6) the conal septum lay to the

Table 1

Pertinent Details of Cases of Single (Primitive) Ventricle

Number	Age	Sex	Situs	Associated abnormalities
			Single (primitive) ventricle with regular (noninverted) transposition in levocardia (normal position of the heart)	
1–216	6 days	M	Solitus; asplenia; mesentery communis	Common atrium; common A-V orifice; straddling aorta with pulmonary atresia; PDA; common left pulmonary vein
2–54	21 wk	M	Solitus; accessory spleen	Subaortic stenosis; right A-V (tricuspid) stenosis; atresia transverse aorta; enlarged pulmonary trunk; PDA; PFO
3–492	21 yr	M	Solitus	Pulmonary stenosis; abnormal pulmonary valve; poststenotic dilatation of pulmonary trunk; PFO; single left and right pulmonary veins; abnormal eustachian valve; hypertrophy and enlargement of right atrium and single ventricle; subacute bacterial endocarditis of right A-V valve; infarct of ventricle
4–2652	1 day	F	Incomplete rotation of stomach; bilobed spleen	Subaortic stenosis; common atrium; common A-V orifice; hypertrophy of single ventricle
5–1916	2½ mo	M	Solitus	Subaortic stenosis; right A-V (tricuspid) valve stenosis; fetal coarctation; PDA; remnants of left venous valve of sinus venosus in right atrium; biatrial hypertrophy and enlargement; hypertrophy of single ventricle; right pulmonary artery banding
6–2330	30 wk	M	Solitus	Subaortic stenosis; fetal coarctation; PFO; PDA; common left pulmonary vein; hypertrophy and enlargement of both atria and the single ventricle
7–375	10 yr	M	Solitus	Subaortic stenosis; PDA; common eustachian and thebesian valve; biatrial and single ventricular hypertrophy
8–512	8 days	F	Solitus	Subaortic stenosis; fetal coarctation; PDA; enlarged pulmonary trunk; PFO; abnormal aortic valve; abnormal band in right atrium; biatrial and ventricular hypertrophy
9–868	2 mo	M	Solitus	Subaortic stenosis; adult coarctation; PDA; right atrial and single ventricle hypertrophy and enlargement; left atrial hypertrophy; banding pulmonary trunk; common left pulmonary vein
10–1989	11 mo	M	Solitus; accessory spleens	Subaortic stenosis; fetal and adult coarctation; common left pulmonary vein; remnants of left venous valve in right atrium; biatrial and single ventricle hypertrophy; right atrial enlargement
11–1206	2 hr	F	Solitus thoracis; spleen in midline; malrotation of intestines	Common A-V orifice with pulmonary atresia; common atrium; LSVC into left side of atrium; absent coronary sinus; PDA; RSVC into right side of atrium; pulmonary veins not identified
12–1872	2 hr	M	Solitus; accessory spleen	Common atrium; common A–V orifice; truncus communis straddling over outlet and main chambers; agenesis of right lung with absent pulmonary artery and vein; common left pulmonary vein

13–436	3 days	F	—	Common atrium; common A-V orifice; pulmonary atresia; right aortic arch; total anomalous pulmonary venous drainage into LSVC into left side of common atrium; right PDA
14	—	—	—	Pulmonary stenosis; PDA
15	—	—	—	Subaortic stenosis; fetal coarctation; PDA; enlarged pulmonary trunk
		Single (primitive) ventricle with inverted transposition in levocardia (normal position of the heart)		
16–595	4 yr 8 mo	F	Solitus	Pulmonary stenosis; right A-V valve stenosis converted to atresia; ASD (fossa ovalis type); biatrial and single ventricle hypertrophy and enlargement
17–225	8 yr 11 mo	M	Solitus	Double left A-V valve; hypoplasia of aorta; enlargement of pulmonary trunk; biatrial hypertrophy; heart block
18–913	6 wk	M	Solitus	Subaortic stenosis; fetal coarctation; PDA; PFO; enlargement of pulmonary trunk; biatrial and single ventricle hypertrophy and enlargement
19–2157	2 mo	F	Solitus	Subaortic stenosis; left A-V valve stenosis; fetal and adult coarctation of aorta; single coronary; common left pulmonary vein; biatrial and single ventricle hypertrophy and enlargement
20–1129	35 yr	F	Solitus	Single coronary; common eustachian and thebesian valve; biatrial and single ventricle hypertrophy
21–1382	4 mo	M	Solitus; bilobed spleen	Left A-V valve stenosis and pulmonary stenosis; PDA; absent eustachian and thebesian valve; right atrial hypertrophy and enlargement; common left pulmonary vein
22–2542	21 days	M	Solitus	Subaortic stenosis; absent transverse arch; PDA; common left pulmonary vein; biatrial and single ventricle hypertrophy and enlargement
23–1274	5½ mo	F	Solitus	Pulmonary stenosis; PDA; PFO; hypertrophy of eustachian valve; right atrial and single ventricle hypertrophy and enlargement
24–1731	15 yr	F	Solitus	Pulmonary stenosis; PFO; common eustachian and thebesian valves; abnormal bands in right atrium; right atrial hypertrophy and enlargement
25–36	22 days	F	Solitus; four lobed left lung	Subaortic stenosis; hypoplasia of the aorta; PFO; PDA; enlargement of pulmonary trunk; absent coronary sinus
26–1768	19 yr	M	Solitus abdominalis; four-lobed left lung; accessory spleen	Subaortic stenosis; left A-V valve stenosis; single coronary; common left pulmonary vein; spontaneous intermittent heart block; right atrial and single ventricle hypertrophy and enlargement; left atrial hypertrophy; parachute left A-V valve.
27–1705	6 days	F	Solitus	Subaortic stenosis; PDA; ASD (fossa ovalis type); aneurysm fossa ovalis; abnormal eustachian valve; biatrial hypertrophy and enlargement; single venricle hypertrophy
28–184	5 yr	M	Solitus	Pulmonary stenosis; bicuspid pulmonic valve; single coronary artery; common left pulmonary vein

Table 1 (continued)

Number	Age	Sex	Situs	Associated abnormalities
29–126	5 yr	M	—	Subaortic stenosis; common conus; common left and right pulmonary vein; hypertrophy of single ventricle
30–2029	10 wk	M	Solitus	Subaortic stenosis; left A-V valve stenosis; coarctation of aorta; PDA; right atrial and single ventricle hypertrophy; left atrial hypertrophy and enlargement
31	—	—	—	Subaortic stenosis; fetal coarctation; ASD (fossa ovalis type); PDA; enlarged pulmonary artery
32	—	—	—	Subaortic stenosis; prominent eustachian valve; common left pulmonary vein
			Single (primitive) ventricle with normal position of arterial trunks in levocardia (normal position of heart)	
33–295	7 yr	F	Solitus	Pulmonary stenosis; ASD (fossa ovalis type); biatrial and single ventricle hypertrophy; left atrial enlargement; left common pulmonary vein
34–674	1 day	M	Solitus	Common A-V orifice; subaortic stenosis; ASD (fossa ovalis type); PDA; bicuspid aortic valve; common eustachian and thebesian valve
35–719	2 mo	M	Solitus abdominalis; trilobed lungs	Common A-V orifice; pulmonary stenosis; right aortic arch; ASD (fossa ovalis type); right atrial hypertrophy; minute left atrium
			Single (primitive) ventricle with transposition, type undetermined, in levocardia (normal position of the heart)	
36–2193	11 mo	M	Solitus; asplenia	Common outlet chamber giving off both trunks, aorta to right, pulmonary trunk to left; common A-V orifice; total anomalous pulmonary venous drainage into right atrium; left atrial atrophy; right atrial and single ventricle hypertrophy; ASD (fossa ovalis type); absent coronary sinus
37–256	Newborn	F	Solitus thoracis; inversus abdominalis	Transposition, type undetermined; aorta anterior; pulmonary trunk posterior; common A-V orifice; pulmonary atresia; total anomalous pulmonary venous drainage into left gastric vein; minute left atrium; right atrial hypertrophy
38–A55–36	1 yr 2 mo	F	Solitus	Transposition, type undetermined; aorta anterior; pulmonary trunk posterior; common A-V orifice; pulmonary stenosis; total anomalous pulmonary venous drainage into SVC; ASD (fossa ovalis type); ASD (coronary sinus type)
			Single (primitive) ventricle in dextrocardia	
39–2688	15 yr	M	Solitus thoracis; no abdominal autopsy	Atria pivotal; complete inverted transposition; common A-V orifice; pulmonary stenosis; bicuspid pulmonic valve
40–2592	2 days	M	Solitus thoracis; no abdominal autopsy	Atria pivotal; inverted transposition with straddling aorta; pulmonary atresia; PDA; ASD (fossa ovalis type)

41–2431	4 yr	M	Solitus	Atrial pivotal; inverted transposition; pulmonary stenosis; unicuspid pulmonary valve; ASD (fossa ovalis type); biatrial and single ventricle hypertrophy
42–17	5 yr 11 mo	M	Solitus thoracis; inversus abdominalis; bilobed spleen	Dextrocardia, type undetermined; inverted transposition; pulmonary atresia; common atrium; common A-V orifice; common left pulmonary vein
43–1337	13 days	F	Inversus thoracis; partial abdominal inversus; asplenia	Dextrocardia, type undetermined; inverted transposition; pulmonary stenosis; common atrium; common A-V orifice; absent coronary sinus
44–87	2 wk	M	Solitus abdominalis; trilobed lungs; asplenia	Subaortic stenosis; dextrocardia, type undetermined; two outlet chambers; regular, (noninverted) transposition; common atrium; common A-V orifice; fetal coarctation; right aortic arch; right PDA
45–1478	5 mo	M	Trilobed lungs; large left lobe of liver, intestinal inversion, with common mesentery; asplenia	Dextrocardia, type undetermined; inverted transposition; total anomalous pulmonary venous drainage into hemiazygos vein; into left-sided atrium; common A-V orifice; absent IVC and coronary sinus; RSVC
			Single (primitive) ventricle in mesocardia	
46–2669	6 days	M	Solitus	Transposition; pulmonary stenosis; bicuspid pulmonic valve; PDA; ASD (fossa ovalis type); juxtaposition of atrial appendages

Abbreviations: PDA = patent ductus arteriosus; PFO = patent foramen ovale; ASD = atrial septal defect; SVC = superior vena cava; LSVC = left superior vena cava; RSVC = right superior vena cava; IVC = inferior vena cava.

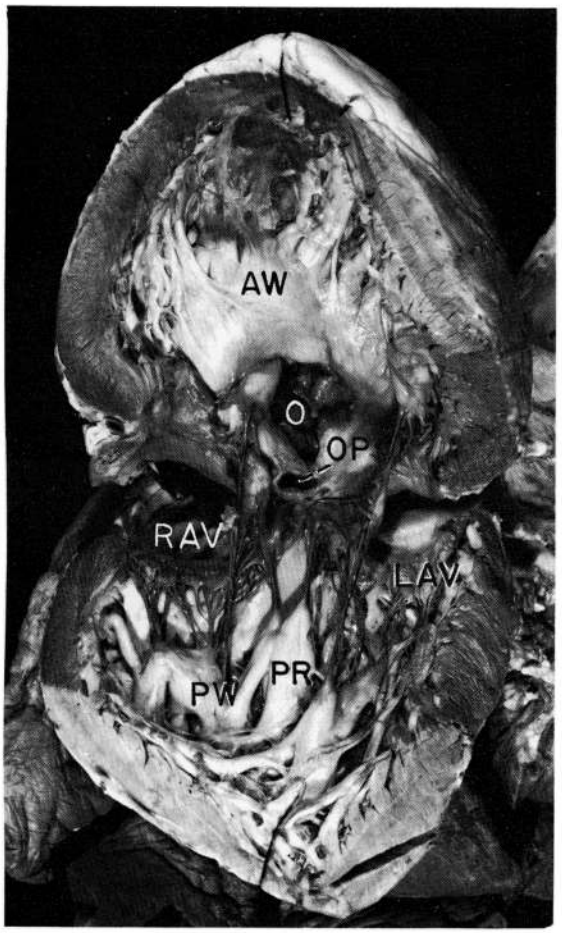

Figure 2

Case 3. Single (primitive) ventricle with regular (noninverted) transposition in levocardia, with pulmonary stenosis. View of single ventricle looking into the base. PW = posterior wall of single ventricle; AW = anterior wall of single ventricle; PR = posterior median ridge; OP = opening into stenotic pulmonary conus; O = opening into small outlet chamber; RAV = right A-V valve; LAV = left A-V valve.

right of the pulmonary trunk; and (7) the anterior papillary muscle of the right A-V valve tended to be attenuated.

The coronary distribution was as follows (fig. 3): The right-sided coronary artery came off the posterior sinus of Valsalva. It gave off the right delimiting branch and continued as the right circumflex artery to supply the right lateral and posterior walls of the ventricle. The left-sided coronary artery came off the left anterior sinus of Valsalva. It gave off the left delimiting coronary artery and then continued as the left circumflex to supply the left lateral and posterior walls of the ventricle.

In this group there were five cases of common A-V orifice and common atrium, two of right A-V valve stenosis, two of pulmonary stenosis, three of pulmonary atresia, nine of subaortic stenosis, and also one case of truncus communis and one of total anomalous pulmonary venous drainage.

Single Ventricle with Inverted Transposition in Levocardia (Normal Position of Heart): 17 Cases (Figs. 4 and 5)

In this type, (1) the outlet chamber was situated to the left and was closer to the left than the right A-V orifice except in three cases, in which it was situated in the middle; (2) the right delimiting coronary artery was more prominent than the left; (3) the left atrial appendage was buried behind the aorta when viewed from the anterior surface of the heart; (4) in most cases, the aorta was situated anteriorly and to the left, and the pulmonary trunk posteriorly and to the right; however, in two cases without pulmonary stenosis and in one with pulmonary stenosis, the aorta was directly anterior to the pulmonary trunk, and in another case with pulmonary stenosis the aorta was anterior and slightly to the right of the pulmonary trunk; (5) the atrial septum had a tendency to be in a more frontal position than in the noninverted type of single ventricle; (6) the conal septum lay to the left of the pulmonary trunk; (7) the anterior papillary muscle of the left A-V valve was attenuated.

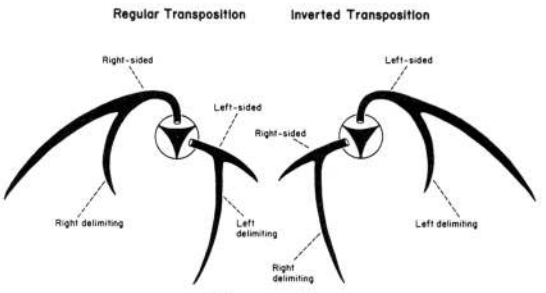

Figure 3

Coronary circulation in single (primitive) ventricle with regular and inverted transposition in levocardia.

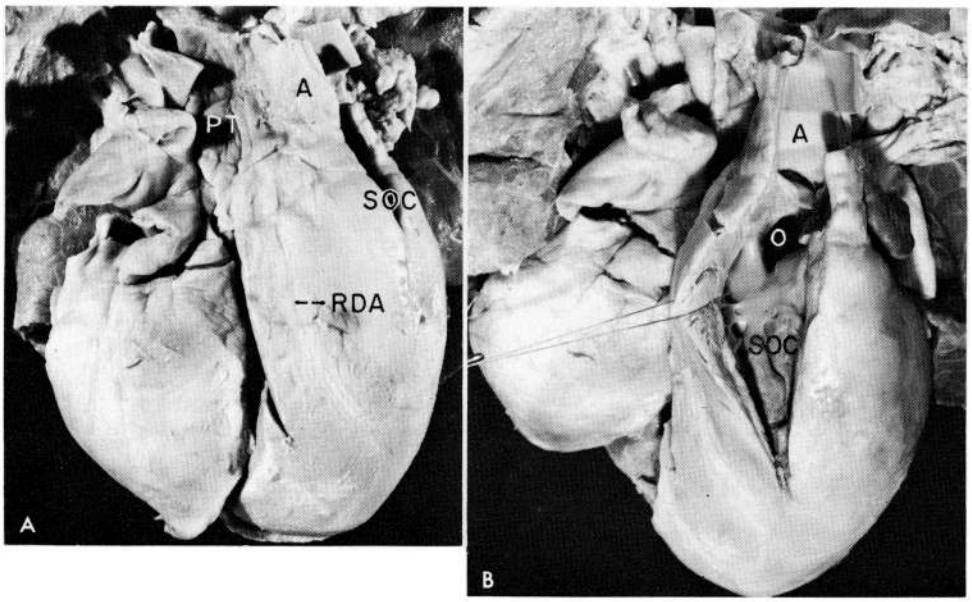

Figure 4

Case 26. Single (primitive) ventricle with inverted transposition in levocardia. (A) Anterior view. (B) View from the left side showing small outlet chamber. See figure 5 for internal area in this heart. RDA = right delimiting artery; A = aorta; PT = pulmonary trunk; SOC = small outlet chamber; O = opening into small outlet chamber with subaortic stenosis.

The origin and distribution of the coronary arteries were mirror-images of those found in the noninverted type when two coronary arteries were present, but a single coronary artery was common. In this group there were four cases of left A-V valvular stenosis, no case of common A-V orifice, one of right A-V valvular stenosis converted to atresia,* five cases of pulmonary stenosis, no case of pulmonary atresia and 10 cases of subaortic stenosis.

In both of the types of single ventricle described (that with regular and that with inverted transposition), the aorta emerged from the small outlet chamber. In one case of regular transposition, there were two outlet chambers each with a vessel arising from it. In both types, situs solitus was generally present, although malrotation of the intestines was found occasionally. The spleen was usually normal, although an occasional case of asplenia, bilobed spleen, or multiple spleens was

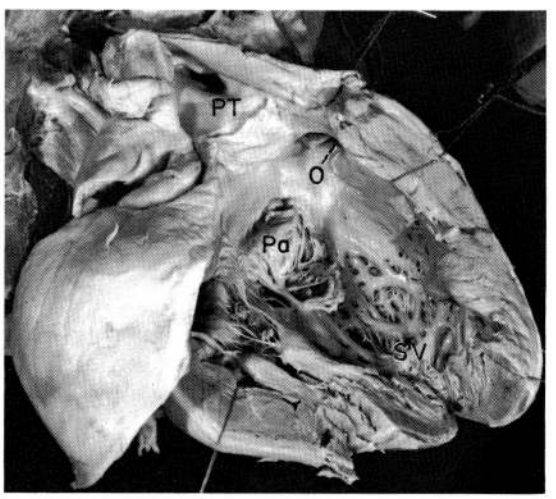

Figure 5

Case 26. Internal view of single ventricle showing stenotic left A-V orifice with parachute valve. O = opening into small outlet chamber with subaortic stenosis; Pa = parachute valve; SV = single ventricle; PT = pulmonary trunk.

*At the gross level an area of a right A-V orifice was filled in by firm tissue. This tissue apparently represented the end stages of thrombosis or reaction to hemodynamic stress, or both. A sinus of the right ventricle was clearly visible.

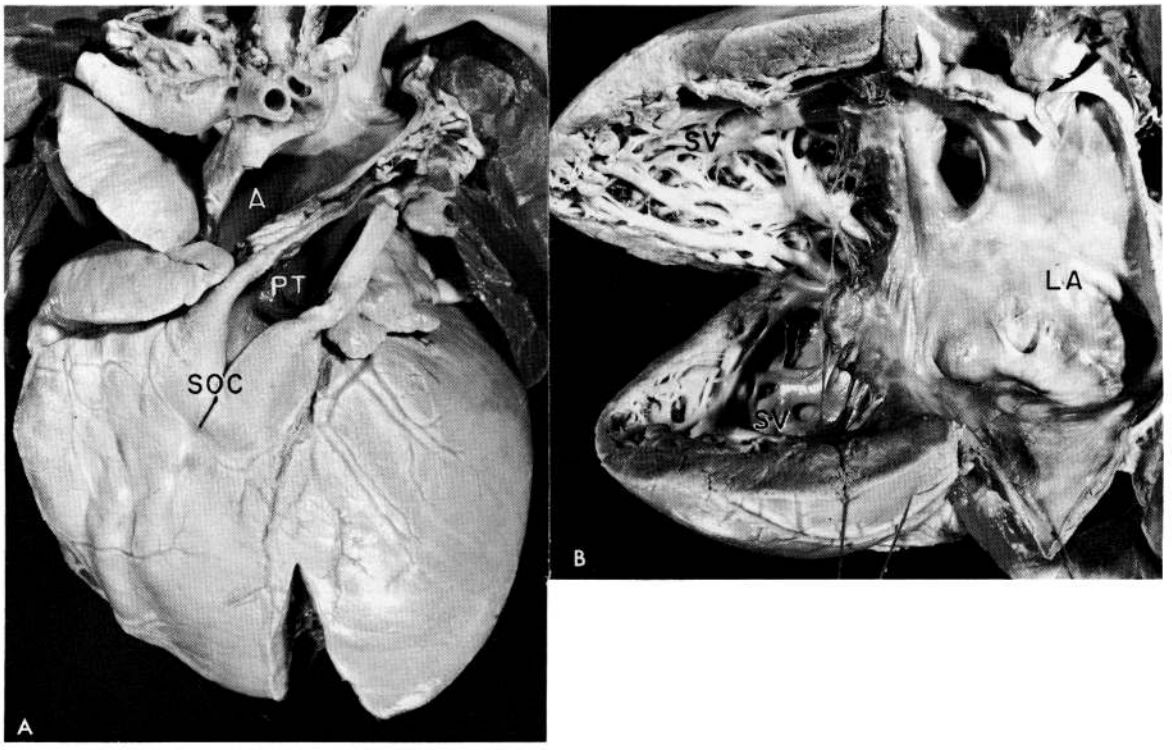

Figure 6

Case 33. Single (primitive) ventricle with normal position of arterial trunks in levocardia, with pulmonary stenosis. (A) Anterior view of the heart. (B) Internal view of single ventricle. A = aorta; PT = pulmonary trunk; SOC = small outlet chamber; LA = left atrium; SV = single ventricle.

noted. Fetal (preductal) or adult (postductal) coarctation was common in both groups. A left common pulmonary vein was noted in many and occasionally, a right common pulmonary vein. In most cases there was biatrial and ventricular hypertrophy.

Single Ventricle with Normal Position of the Arterial Trunks in Levocardia (Normal Position of Heart), Holmes Heart: Three Cases (Figs. 6 and 7)

In this type (1) the pulmonary trunk emerged from the small outlet chamber; (2) the aortic annulus was continuous with the left A-V or common A-V annulus. In one case both mitral and tricuspid valves were present (fig. 6); in two, a common A-V orifice was observed (fig. 7). In two, there was pulmonary infundibular stenosis, and in one, subaortic stenosis. In the one case with good survival (7 years), biatrial and ventricular hypertrophy were observed.

Single (Primitive) Ventricle with Transposition, Type Undetermined, in Levocardia (Normal Position of Heart): Three Cases

In this group, although the diagnosis of single ventricle was clear according to our definition, it was not possible to classify the type of transposition because of insufficient criteria; that is, the relationship of the arterial trunks to each other, the position of the outlet chamber, and the size of the delimiting arteries were not sufficiently distinctive to permit a diagnosis. All three hearts had a common A-V orifice and total anomalous pulmonary venous drainage. One had associated pulmonary stenosis.

There were three cases (in levocardia) which were not included in the present series because of uncertainty as to whether they belonged to the category of single ventricle. In this group the outlet chamber was elongated and extended inferiorly down to the acute margin. The extended area possessed no

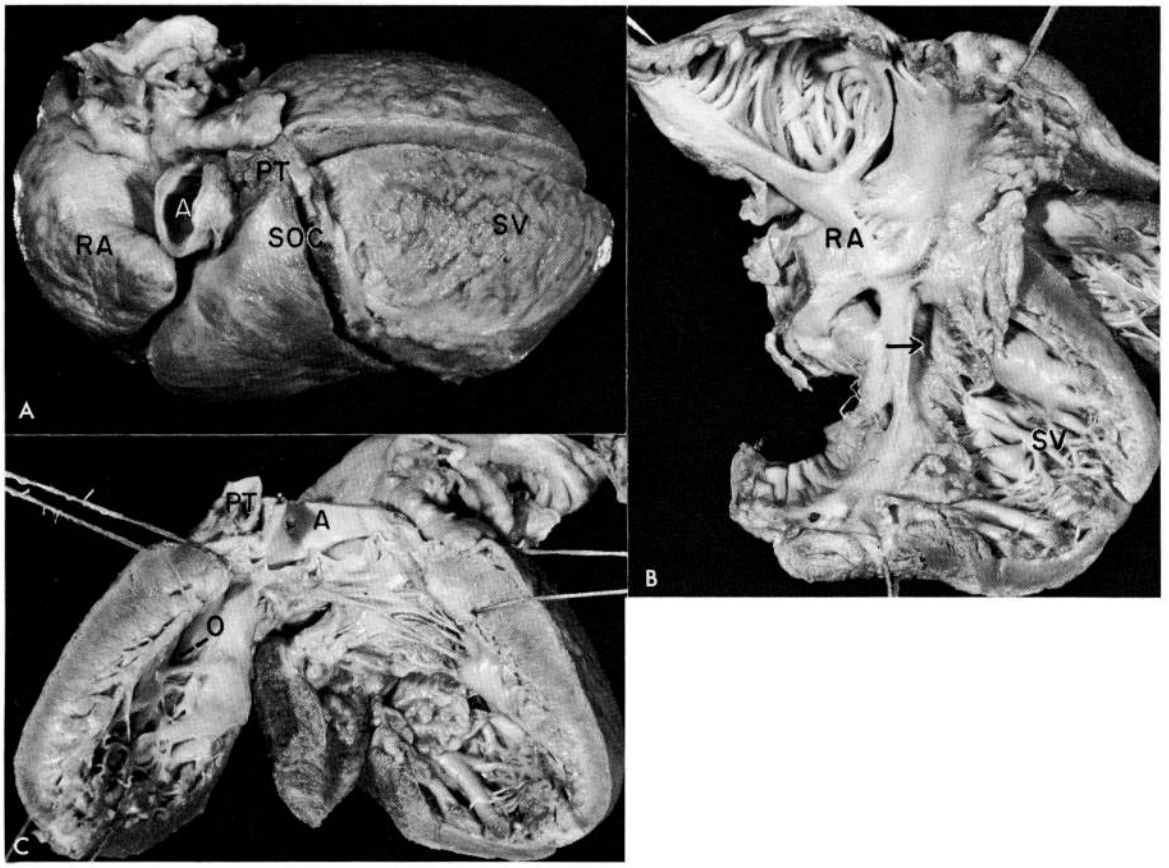

Figure 7

Case 35. Single (primitive) ventricle with normal position of arterial trunks in levocardia, with common A-V orifice and pulmonary stenosis. (A) Antero-superior view of the heart. (B) View of right atrium and single ventricle. (C) View of single ventricle and aorta. A = aorta; PT = pulmonary trunk; SOC = small outlet chamber; RA = right atrium; SV = single ventricle; O = opening into small outlet chamber. Arrow points to opening of left atrium into single ventricle.

papillary muscles, but the lower part of the ventricular septum between the two chambers could be interpreted as a posterior ventricular septum. Therefore, although the mitral and tricuspid orifices entered the larger chamber, it was difficult to ascertain whether or not the small chamber had a sinus component in it.

Single Ventricle in Dextrocardia: Seven Cases

In three cases the atria were in pivotal position[4] (left atrium to the left and anterior, right atrium to the right and posterior), and inverted transposition was associated with pulmonary stenosis or atresia. In these cases there was situs solitus of the abdominal viscera. In the other four cases, asplenia with undecided situs dominated the picture associated with common atrium, common A-V orifice, pulmonary stenosis or atresia. In three of these latter four cases there was inverted transposition, and in one regular noninverted transposition.

Single Ventricle in Mesocardia: One Case

In the one case in this series, there was regular noninverted transposition with pulmonary stenosis.

Differential Diagnosis of Single Ventricle from Allied Conditions

Common Ventricle

In common ventricle, as we refer to it, there is no common ventricular sinus. Instead, the

ventricular chamber is clearly subdivided by a portion of the true posterior ventricular septum into a right and a left sinus. Each sinus is related to a separate conus. We are thus dealing with a huge ventricular septal defect with distinct right and left ventricular sinus components.

Straddling Tricuspid Orifice

In this entity[5,6] the basic anomaly is mitral atresia or mitral stenosis, with the common A-V canal type of ventricular septal defect, or the latter by itself. Here the small right-sided chamber can be seen to have a sinus as judged by the papillary muscle attachments. The same is true of the dominant left ventricular type of common A-V orifice. The right-sided chamber is a true right ventricle, since papillary muscles are present in the abbreviated sinus.

Tricuspid and Mitral Atresia Complexes

These may be ruled out by our definition which requires that both A-V orifices or a common A-V orifice must enter a common ventricular sinus. A question arises, however, as to whether in these anomalies a remnant of ventricular sinus is present, hidden in the opposite sinus, related to the atretic A-V orifice. If this is so, these anomalies might be placed in the category of single ventricle. This is not the case in tricuspid atresia without transposition. Studies of the conduction system by one of us (M. L.)[7] have shown a right bundle branch separating a distinct sinus from a conus of the right ventricle, and hence there are two distinct ventricles in this anomaly. Neither is this the case in mitral atresia which is a part of hypoplasia of the aortic tract complex,[8] nor in mitral atresia with ventricular septal defect with or without transposition,[9] in which two distinct ventricles separated by an anterior (bulbar) and posterior (main) portion of the ventricular septum are evident grossly. In tricuspid atresia with transposition (ordinary or inverted), however, the possibility remains that a sinus of the right ventricle may be hidden in the left. Further studies of the conduction system will be helpful here.

Discussion

Hearts with single and common ventricles, as defined above, were described as early as 100 years ago.[10–22] However, various other types were included in these categories, such as hypoplasia of the aortic tract complex, which do not fit our definitions. The more recent literature has been reviewed by Campbell and associates,[23] Edwards,[24] Van Praagh's group[25] and Elliott and co-workers.[26]

Our concept of single ventricle is, in general, in keeping with that of Taussig,[27] Brown,[28] Rogers and Edwards,[29] Harley,[30] Campbell and associates,[23] Edwards,[24] and Elliott's group.[26] However, we distinguish between single and common ventricle, which was not done by the other authors, and exclude from these categories mitral and tricuspid atresia, which were included by some of the other authors.

Van Praagh and associates[25] defined their concept of single ventricle according to the absence of the right or left sinus or both ventricular sinuses, or of the sinus portion of the ventricular septum, as well as according to the type of bulboventricular loop. We could not base the diagnosis in our cases on the absence of a sinus, since we could not determine whether the walls of the common sinus seen in our cases of single ventricle belonged structurally to the left or the right ventricle. Our cases of single ventricle correspond morphologically to type A of Van Praagh and co-workers, and our common ventricle corresponds to their type with absence of the sinus portion of the ventricular septum.

Recently, de la Cruz and Miller[5] have attempted to alter the entire concept of single ventricle as we have defined it. They include the type of heart described in this paper as single ventricle in the category of *double-inlet left ventricle,* thus excluding it from what they interchangeably call single ventricle, common ventricle, and cor biatriatum triloculare. They apply the latter three terms to that entity in which the interventricular septum is absent or is represented by a small muscular ridge, with the atria entering into and the great vessels

arising from a single chamber. This corresponds to our term "common ventricle."

This alternate concept of double-inlet left ventricle is based on an embryological concept. In the second phase of the development of the heart, as the bulbus shifts to the left and is absorbed into the ventricles, the auricular canal shifts to the right and this shift makes possible the expansion of the tricuspid orifice and the elaboration of the sinus of the right ventricle.[31] According to the concept of de la Cruz and Miller[5] and Goerttler,[32] varying degrees of lack of shift result in either complete placement of both A-V orifices into the primitive ventricle (left ventricle of de la Cruz) or a straddling tricuspid orifice entering into both chambers. Following this concept, some of the hearts excluded from single ventricle by us (straddling tricuspid orifice) are included in the category of double-inlet left ventricle. Whether one should adopt the concept of de la Cruz and Miller or espouse the one outlined in this paper is a matter of taste in selecting an embryological or an anatomic basis for classification.

From all points of view (Harley,[30] de la Cruz and Miller,[5] and Goerttler[32]), the embryological abnormality in single ventricle, as defined by us, or double-inlet left ventricle may be considered to be persistence of the primitive state of the bulboventricular loop, whether inverted or noninverted, due fundamentally to a lack of expansion and shift of the atrial canal to the right (or left, when inverted) during the process of absorption of the bulbus into the heart.

The question of the identity of the main ventricular chamber arises in what we call single ventricle. Although Van Praagh and associates[25] and de la Cruz and Miller[5] considered this the left ventricle, we were unable to identify the main ventricular chamber from the morphology alone. Nor is embryological analysis helpful, for opinions differ as to the fate of the descending and ascending limbs of the bulboventricular loop. De la Cruz and Miller,[5] quoting Streeter, are of the opinion that the descending limb gives rise to the sinus of the left ventricle and the ascending limb to the entire right ventricle and the conus of the left ventricle. Pernkopf and Wirtinger[33] and Goerttler[31] stated that the descending limb (proampulla) gives rise to the sinuses of both ventricles and the ascending limb (metampulla and bulbus) to the coni of both ventricles and in part to the sinus of the right ventricle. If the former opinion is correct, then the single ventricle is the definitive left ventricle and the outlet chamber is the conus of the right ventricle. If the latter is true, then the single ventricle is not the definitive left ventricle but includes in it a portion of sinus of the right ventricle. Because of this difference of opinion, it may be preferable to identify the main ventricular chamber as a primitive ventricle as Campbell and co-workers,[23] Goerttler,[32] and Harley[30] have done.

The nature of the septum between the main and outlet chamber in single ventricle, discussed extensively by Harley[30] and Van Praagh's group,[25] is related to the problem of the nature of the main ventricular chamber. The elucidation of both of these problems awaits studies of the conduction system.

In our diagnosis of the type of transposition in single ventricle, we used a summation of criteria rather than the position of arterial trunks alone. These criteria include in addition to the position of the arterial trunks, the position of the outlet chamber and its relation to the A-V orifices, the relative size of the delimiting coronary arteries, and the position of the conus septum vis-a-vis the pulmonary trunk or aorta. When the diagnosis had been made on the basis of all of these criteria, it became evident that if the aorta was frankly to the left and the pulmonary trunk frankly to the right, then there was always inverted transposition. If the aorta was frankly to the right and anterior and the pulmonary trunk frankly to the left and posterior, then there was always ordinary (noninverted) transposition. If, however, the aorta was directly anterior, or anterior and only slightly to the left or right, then the type of transposition was difficult to classify from the position of the vessels alone, and only a summation of criteria

made the diagnosis possible. In some cases the criteria were inadequate for a diagnosis, and hence these hearts were categorized as of undetermined type. The findings concerning the position of arterial trunks mirror those of Elliott and co-workers.[26]

The presence in this series of three hearts, which may be designated as the Holmes type of hearts, adds to the series reviewed by Rosenquist and associates,[34] and Elliott's group.[35] We are including cases with common A-V orifice in the concept of the Holmes heart.

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