Isolated ventricular inversion with situs solitus

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The clinical and anatomical findings in two patients with isolated ventricular inversion and situs solitus are described. The other 4 previously published cases are reviewed. The 6 patients with this malformation, all without pulmonary stenosis, presented a clinical picture of cyanotic congenital heart disease, associated with increased pulmonary blood flow (hypoxaemia and cardiac failure). The importance of different diagnostic tests is discussed and it is concluded that angiocardiology is the only definitive means of establishing the diagnosis. Because the physiopathological disturbance is the same as in transposition of the great arteries, both malformations should be similarly considered with respect to diagnosis and treatment. Nevertheless, the high incidence of certain associated malformations in cases of isolated ventricular inversion adds to difficulty in diagnosis, and makes a good result from the Mustard procedure less likely than in transposition of the great arteries.

In 1966 Van Praagh and Van Praagh described a malformation characterized by ventricular inversion with situs solitus of the viscera and atria and normally related great arteries, using for the first time the term 'isolated ventricular inversion'.

The course of the circulation in this cardiac anomaly is as follows: the aorta receives venous blood from the right atrium and the venae cavae through a right-sided morphological left ventricle; the pulmonary artery receives arterial blood from the pulmonary veins and left atrium through the left-sided right ventricle (Fig. 1). As a result, this entity clinically resembles complete transposition of the great arteries: the description 'clinical complete transposition of the great arteries with situs solitus', was used by Stanger *et al.* (1968). Similar malformations had been reported previously by Lev and Rowlatt (1961) and by Ratner, Abbott, and Beattie (1931). Only 2 cases of this malformation have been reported since Van Praagh's description, by Martínez Picó and Muñoz (1967) and Pérez Treviño, Hurtado del Río, and Mae Holden (1972).

The purpose of this paper is to add to knowledge of this entity, by reporting 2 more cases of isolated ventricular inversion with situs solitus. Attention is called to some diagnostic and therapeutic difficulties related to the anatomical findings.

In order to understand the type of conal alterations present in the cases reported in this paper, we define the terms subaortic conus, subpulmonary

conus, conal septum, and conal free wall, as used in this paper.

When present, the *subaortic* and *subpulmonary coni* are complete muscular rings consisting of two components: the conal septum (the component common to both coni) and the subaortic and subpulmonary conal free walls (specific components relating to the subaortic and subpulmonary conus, respectively).

The term *conal septum* is given to the muscular septum separating the subaortic from the subpulmonary outflow tract. It lies beneath the coronary aortic sinuses and, when the ventricular septum is viewed from its left ventricular aspect, that part derived from the conal septum may be delimited by imaginary lines running vertically through the midpoints of the coronary leaflets of the aortic valve. The term *conal free wall* (subaortic or subpulmonary) will be reserved for the remainder of the subaortic or subpulmonary conus. If there is continuity between an atrioventricular valve and a semilunar valve it is because of absence of the conal free wall rather than the conal septum.

As well as reporting our two cases, we discuss the main anatomical and clinical characteristics of the cases previously reported.

Case reports

Case 1

A 7 month-old female infant (Case 5 of Tables 1, 2, and 3) was admitted to the hospital because of cyanosis and

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FIG. 1 Course of the circulation in isolated ventricular inversion with situs solitus of the viscera and the atria. Note how the aorta (Ao) receives venous blood coming from the venae cavae (IVC and SVC), the right atrium (RA), and the right-sided morphological left ventricle (LV). The pulmonary artery (PA) receives arterial blood from the pulmonary veins (PV), the left atrium (LA), and the left-sided morphological right ventricle (RV). The main physiopathological disturbance is, therefore, similar to that observed in complete transposition of the great arteries.

dyspnoea since birth. Underdevelopment, generalized cyanosis, increased praecordial activity, a liver edge 3 cm below the right rib margin, and a grade 2/6 systolic murmur were the salient findings at the physical examination.

Right atrial and ventricular enlargement were present on the electrocardiogram (Fig. 2 A).

Chest x-rays revealed cardiomegaly, a very prominent right heart border, and increased pulmonary circulation probably caused by both pulmonary venous congestion and pulmonary plethora (Fig. 3A).

The following findings were obtained at cardiac catheterization and angiocardigraphy: 1) Normal location and drainage of the great veins (pulmonary and systemic) and normal location of the atria, as assessed

by normal position and drainage of the great veins and direct visualization of the atria themselves (Fig. 4A and 4B). 2) Two ventricles, one right-sided with the anatomical features of the left ventricular chamber, were separated by a large filling defect interpreted as the conal septum; the ventricles seemed to be connected by 2 apparent ventricular septal defects situated above and below the large conal septum (Fig. 4A). 3) The aorta emerged from the right-sided morphological left ventricle to the right (Fig. 4A) and behind (Fig. 4C) the pulmonary artery which took its origin from the left-sided morphological right ventricle (Fig. 4A and 4B). The left atrium was filled (Fig. 4B) after injection of contrast material into the pulmonary artery; it emptied slowly into the left-sided ventricle. 5) The pulmonary trunk and the persistent ductus arteriosus were catheterized through the superior ventricular septal defect. The pulmonary artery pressure was at systemic level.

Course A Blalock-Hanlon operation and banding of the pulmonary artery were indicated. The patient died in irreversible congestive heart failure 3 days after the Blalock-Hanlon operation.

Pathological findings Both atria and their tributary veins were topographically and morphologically normal. The foramen ovale was sealed. The apex pointed to the left. The aorta was located to the right and posteriorly with respect to the pulmonary artery. There was a left aortic arch. The right atrium opened through a mitral valve into a right-sided morphological left ventricle (Fig. 5 and 6). The dimensions of the mitral orifice were 20 mm long and 13 mm wide. The distance of the mitral annulus from the interior of the apex of the left ventricle was 40 mm. The aorta arose from this ventricle, with fibrous continuity between aortic and mitral valves (Fig. 5 and 6), because of a lack of a subaortic conal free wall. There was a persistent ductus arteriosus.

The left atrium opened into a left-sided morphological right ventricle, smaller than the left one, through a tricuspid valve. The dimensions of the tricuspid orifice were 12 mm long and 6 mm wide. 1 to 2 mm above the tricuspid valve there was a stenotic ring. The distance from the tricuspid annulus to the interior of the apex of the right ventricle was 18 mm. The right ventricular cavity was, therefore, significantly smaller than the left. The ventricular septum was underdeveloped, being reduced to its posterior and apical portions. The lack of membranous and anterior components (the septal band was absent) gave rise to a huge, anterior ventricular septal defect (Fig. 6). The hypertrophied conal septum joined, posteriorly, the anterior border of the posterior portion of the ventricular septum (Fig. 6). The superior border was continuous with the truncal and arterial septa (Fig. 6). The anterior portions of both (the inferior border of the conal septum and the superior border of the apical remnant of the ventricular septum) diverged as they proceeded in an anterior direction, giving rise to an outlet structure connecting the left ventricle with a small portion of the pulmonary artery which is above that cavity as a result of the anomalous rightward shifting of the conal septum (Fig. 6).

In view of these facts, the left ventricular angiocardio-

FIG. 2 a) Electrocardiogram of Case 1 (Case 5 in Tables 1, 2, and 3) suggests enlargement of the right atrium and ventricle. b) Electrocardiogram of Case 2 (Case 6 in Tables 1, 2, and 3) revealing right atrial and right ventricular enlargement.

gram was reviewed and a retrospective interpretation, based upon the anatomical information, was made. What had been considered to be a ventricular septal defect below the conal septum was now interpreted as this orifice contained in the right-sided left ventricular cavity, between the conal and ventricular septa (Fig. 4A, 5, and 6). The apical remnant of the ventricular septum was thought to correspond to the left lowest contour of the left ventricular cavity (Fig. 4A). The dashed line would correspond to the anterior large ven-

tricular septal defect, the posterior smaller one occupying the place given in the previous interpretation (Fig. 4A).

Case 2

This patient, a 5-month-old girl, (Case 6 of Tables 1, 2, and 3) had been cyanosed and dyspnoeic from birth. On examination she was deeply cyanosed. Peripheral pulses were regular. Palpation of the praecordium showed an enlarged heart. A grade 2/6 systolic murmur

TABLE I *Clinical features*

Author	Case No.	Age	Sex	Clinical features	Radiology			
					Apex	Gastric bubble	Configuration of heart shadow	Pulmonary circulation
Lev and Rowlatt (1961)	1	1 mth	M	Hypoxaemia ++ Congestive heart failure ++	Left	—	Prominent pulmonary artery	
Van Praagh and Van Praagh (1966)	2	4 mth	F	Hypoxaemia ++ Congestive heart failure ++	Left	Left	Cardiomegaly; left atrial enlargement; prominent pulmonary artery	↑
Martínez Picó and Muñoz (1967)	3	3 mth	F	Hypoxaemia ++ Congestive heart failure	Left	Left	Cardiomegaly	↑
Pérez Treviño <i>et al.</i> (1972)	4	12 dy	M	Hypoxaemia ++ Congestive heart failure +++	Left	Left	Cardiomegaly	↑
Present cases (1974)	5	7 mth	F	Hypoxaemia ++ Congestive heart failure ++	Left	Left	Cardiomegaly; right atrial enlargement	↑
	6	5 mth	F	Hypoxaemia ++ Congestive heart failure ++	Left	Left	Cardiomegaly; right atrial enlargement; prominent pulmonary artery	↑

↑: increased;

FIG. 3 *a*) Case 1 (Case 5 in Tables 1, 2, and 3). Chest x-ray, frontal projection, shows an unusually prominent right cardiac border, cardiomegaly, and increased pulmonary circulation, probably caused both by pulmonary congestion and plethora. *b*) Case 2 (Case 6 in Tables 1, 2, and 3). Similar x-ray findings.

Electrocardiogram								Follow-up and surgical result
QRS axis	aVR	V ₃ R	V ₁	V ₆	Ventricular hypertrophy	Atrial hypertrophy	P wave	
-48°	QS	RS	rS	RS	Both	Both	+ in I; - in aVR	Banding; death 2 days later during bronchoscopy
+120°	rSr'							
+70°	Qr		rS (T+)	Rs (T-)	Left	Both		Blalock-Hanlon proposed, but not performed
+140°	qR	Rs	Rs	rS	Right	Right	+ in I - in aVR	No surgery; death 2 hr after cardiac catheterization, from cardiac arrest
+140°	qR		R	Rs	Right	Right	+ in I; - in aVR	Blalock-Hanlon; death 3 dy after operation
								No surgery; death 3 dy after admission

was heard. The liver edge was 2 cm below the right costal margin.

The electrocardiogram (Fig. 2B) revealed a P axis of +60°. The P wave was peaked and high suggesting right atrial enlargement. The QRS axis was +140°. Tall R waves in the right praecordial leads and aVR suggested right ventricular hypertrophy.

Chest x-ray (Fig. 3B) revealed cardiac enlargement, pulmonary plethora, and a prominent right cardiac border.

Cardiac catheterization and angiocardiogram in the Hospital de la Cruz Roja, Barcelona, Spain, were interpreted as showing transposition of the great arteries.

Course The patient died 3 days after admission, with cardiac failure and hypoxaemia.

Pathological findings There was situs solitus of the viscera and atria. The apex pointed to the left. The aorta was located to the right and slightly behind the pulmonary artery. There was a left aortic arch. The ductus arteriosus was patent. The right atrium opened through a normal mitral valve into an enlarged, morphological left ventricle (Fig. 7). The dimensions of the mitral orifice were 18 mm long and 10 mm wide, the distance from its annulus to the interior of the left ventricular apex being 38 mm. The aorta arose from this ventricle with fibrous continuity between aortic and mitral valves (Fig. 7). The aortic valve was separated from a sub-aortic ventricular septal defect by a muscular flange (Fig. 7). There was right juxtaposition of the atrial

appendages. The left atrium opened through a small tricuspid valve into a hypoplastic morphological right ventricle (Fig. 8). The dimensions of the tricuspid valve were 5 mm long and 5 mm wide, the distance from its annulus to the interior of the apex of the right ventricle being only 14 mm. A comparison of the ventricular dimensions confirmed the impression that the right ventricle was much smaller than the left. Above the tricuspid valve a stenotic fibrous ring existed. The ventricular septal defect observed from the right ventricle seems to be the result of a malalignment between the conal septum and the septal band, the former being displaced leftwards and forwards (Fig. 8), and the ventricular septal defect being located above the superior border of the septal band, between the anterior and posterior divisions of this last structure (Fig. 8).

Discussion

The malformation described under the name of isolated ventricular inversion (Van Praagh and Van Praagh, 1966) consists of a ventricular inversion with situs solitus of the viscera and atria and normal position of the great arteries (Fig. 1). The 2 cases described in this paper and 4 others previously reported (Lev and Rowlatt, 1961; Van Praagh and Van Praagh, 1966; Martínez Picó and Muñoz, 1967; Pérez Treviño *et al.*, 1972) fulfil this definition.

TABLE 2 Findings at cardiac catheterization and angiocardiology

Author	Case No.	Anatomical right atrium	Atrial septal defect	Anatomical left atrium	Anatomical right ventricle	Anatomical left ventricle	Ventricular septal defect	Inferior vena cava	Superior vena cava
Lev and Rowlatt (1961)	1	Not performed							
Van Praagh and Van Praagh (1966)	2	Right-sided			Left-sided?	Right-sided	One high, sub-aortic; one low, muscular	Right	Right
Martínez Picó and Muñoz (1967)	3	Right-sided				Right-sided; anterior			
Pérez Treviño <i>et al.</i> (1972)	4	Right-sided	Sealed	Not visualized	Not visualized	Right-sided	None	Right	Right
Present cases (1974)	5	Right-sided	Sealed	Visualized by injection in pulmonary artery, slow emptying into left ventricle	Left-sided	Right-sided	One high, subaortic; one low (see text and Fig. 5)	Right	Right
	6	Data not available

a

b

Great arteries	Pressures (mmHg)	Oxygen satn (%)
Aortic valve low and posterior to pulmonary artery; aorta originating from right-sided left ventricle; larger pulmonary artery from left-sided right ventricle	Descending aorta 60/20 Ascending aorta Anatomical right ventricle 60/0-10 Right atrium 5/-5 Superior vena cava Inferior vena cava	83 69 79 59 39 37
Aorta arising from right-sided left ventricle		
Aorta to right originating from right-sided left ventricle		
Aorta arising from right-sided anatomical left ventricle; aorta to right and posterior to pulmonary artery, the latter arising from left-sided right ventricle	Superior vena cava Inferior vena cava Right atrium 10/0-6 Anatomical left ventricle 70/2 Anatomical right ventricle 65/10 Ascending aorta 60/30 Descending aorta Pulmonary artery 56/30	69 62 66 70 90 67 60 74

c) Contrast injection into the morphological left ventricle (LV) in the lateral projection. Note the aorta (Ao) posterior to the pulmonary artery (PA).

FIG. 4 Angiocardiographic findings in Case 1 (Case 5 in Tables 1, 2, and 3). a) Contrast injection into the right-sided morphological left ventricle (LV) from which the aorta (Ao) originates and proceeds to the right of the pulmonary artery (PA). Note the smooth contours and triangular shape of this cavity. There is contrast material regurgitating from this chamber into another one which is presumed to be a normally situated right atrium because it is receiving a right-sided inferior vena cava. The apical portion of the ventricular septum (VS) separates the right-sided left ventricular cavity from the remainder of the left portion of the heart silhouette, which will prove to be the left-sided right ventricle (Fig. 4B, RV). The large filling defect localized between the subpulmonary (SPOT) and subaortic (SAOT) outflow tracts is thought to be the conal septum (CS) abnormally hypertrophied and displaced towards the right-sided morphological left ventricle. Between the apical portion of the ventricular septum (VS) and the conal septum (CS) there is an orifice (arrow) through which contrast material from the left ventricle passes into the pulmonary artery (PA) which, owing to the rightward shifting of the conal septum (CS) overrides, to some degree, the ventricular septum (VS). (The reasons for believing that this hole between the conal and the ventricular septa is not a muscular ventricular septal defect have been discussed in the text.) Perforating the upper part of the conal septum (CS) just below its junction with the truncal septum (TS), there is a small ventricular septal defect (VSD) related to both great arteries. The large ventricular septal defect involving the membranous septum and an extensive anterior portion of the muscular septum is indicated by the dotted line which has been drawn between the upper end of the apical ventricular septum and the point of junction of the conal (CS) and truncal (TS) septa.

b) Contrast injection to the pulmonary artery showing the normally situated left atrium (LA) with its normal, rounded, smooth appearance, the right ventricle (RV), and the pulmonary artery (PA).

FIG. 5 Anatomical findings. Case 1 (Case 5 in Tables 1, 2, and 3). Internal view of the right-sided left ventricle. Observe the aortic valve (Ao) in fibrous continuity (*) with the anterior mitral leaflet (MV). Note the ventricular septal defect (VSD), and the orifice limited by a hypertrophied conal septum (CS) to the right, and the ventricular septum (VS) to the left. This hole has an area which is parallel to the horizontal plane and is completely located in the left ventricular cavity, to the right of the ventricular septum (VS). It does not connect the two ventricles, being only the entrance to an outflow tract going to the pulmonary artery.

FIG. 6 Diagrammatic representation of the anatomical features of Case 1, showing the unusual relation of the great arteries, the conus cordis, and the ventricles. For the sake of clarity, the atria and their tributary veins (entirely normal, both from the anatomical and topographical view-points) have been omitted from the diagram. The drawing appears as if the anterior right and left ventricular free walls and the anterior portion of the conal (CS) and ventricular (VS) septa had been excised. The morphological left ventricle (LV) is a right-sided cavity from which the aorta (Ao) arises. The aortic semilunar leaflets (AoV) are in fibrous continuity (MAOC) with the aortic leaflet of the mitral valve (MV), in the absence of a subaortic conal free wall.

The morphological right ventricle (RV), considerably smaller than the left, is a left-sided cavity. The pulmonary artery (PA) originated from this cavity, and is supported by a subpulmonary conal free wall (SPCFW), so that there is no fibrous continuity between the pulmonary (PV) and tricuspid (TV) leaflets. The ventricular septum (VS) was very underdeveloped. The lack of the membranous and anterior components (the septal band is absent and the conal septum is shifted to the right) gave rise to a very large ventricular septal defect, represented in the drawing by a dashed line (DL) going from the superior border of the ventricular septum (VS) to the point of junction between both the conal (CS) and truncal (TS) septa. The conal septum (CS) is very enlarged and displaced towards the right in such a way that it is really contained in the right-sided morphological left ventricle (LV). Between the conal (CS) and ventricular (VS) septa a funnel-like passage is formed which leads from the left ventricle (LV) to the pulmonary artery (PA) (follow arrow 1 a) and through the ventricular septal defect described above to the morphological right ventricle (RV) (follow arrow 1 b). The entrance to this funnel-like structure (see beginning of arrow 1 a and 1 b) has an ellipsoid shape, with its plane parallel to the horizontal. It is completely contained in the morphological right-sided left ventricle (LV) to the right of the ventricular septum and may be mistaken for a muscular ventricular septal defect. The posterior end of the conal septum (CS) fuses with anterior border of the posterior muscular ventricular septum (dotted line) (dl). Its anterior extremity fuses with the anterior free wall of the morphological left ventricle (LV). Since, in this drawing, all the anterior wall of the heart has been removed, the most anterior part of the conus seen is a cross-section of its anterior portion. The fusion of this portion of the conal septum (CS) with the anterior free wall of the morphological left ventricle is seen in Fig. 5. Some millimetres below the junction between the truncal (TS) and conal (CS) septa there is another funnel-like structure connecting the ventricles at the level of their outflow tracts (arrow 2).

The clinical and anatomical features of the 2 cases described here and of those already published are summarized in Tables 1, 2, and 3.

The age at death varied between 12 days and 7 months. There were 2 male and 4 female patients. The clinical picture was characterized by congestive heart failure and hypoxaemia in all cases (as in transposition of the great arteries without pulmonary stenosis).

The cardiac apex pointed to the left in 4 cases. In Cases 5 and 6, an unusually prominent right heart border prevented us from locating the exact position of the cardiac apex. There was a bulging pulmonary artery segment in Cases 1 and 2 and left

atrial enlargement in Case 2. Pulmonary plethora and cardiomegaly were constantly present. The unusually prominent right heart border in Case 6 (Fig. 3B) may have been caused by the right juxtaposition of the atrial appendages.

The electrocardiogram revealed a frontal QRS axis ranging between -48° and $+140^\circ$. No patient had an electrocardiogram strongly suggesting a diagnosis of ventricular inversion. The right ventricular hypertrophy shown by the electrocardiogram of Cases 5 and 6 (Table 1, Fig. 2A and 2B) is considered to be the result of relative hypoplasia of the left-sided morphological right ventricle.

The haemodynamic and angiographic find-

TABLE 3 *Anatomical findings*

<i>Author</i>	<i>Case No.</i>	<i>Visceral and atrial position</i>	<i>Apex</i>	<i>Bulboventricular loop</i>	<i>Great arteries position</i>	<i>Aortic arch</i>	<i>Juxta-position of atrial appendages</i>
Lev and Rowlatt (1961)	1		Left	L-loop	Aorta to right of pulmonary artery	Left	
Van Praagh and Van Praagh (1966)	2	Solitus	Left	L-loop (morphological left ventricle anterior and to right of morphological right ventricle)	Aorta posterior and to right of pulmonary artery	Left	
Martínez Picó, and Muñoz (1967)	3	Solitus	Left	L-loop	Aorta posterior and to right of pulmonary artery	Left	
Pérez Treviño <i>et al.</i> (1972)	4	Solitus	Left	L-loop	Aorta posterior and to right of pulmonary artery	Left	
Present cases (1974)	5	Solitus	Left	L-loop	Aorta posterior and to right of pulmonary artery	Left	
	6	Solitus	Left	L-loop	Aorta posterior and to right of pulmonary artery	Left	Right-sided

ings in all the cases in which these investigations were performed were: 1) situs solitus of the viscera and atria; 2) a right-sided morphological left ventricle giving rise to the aorta, situated behind and to the right of the pulmonary artery; 3) left-sided morphological right ventricle from which the pulmonary artery emerged (Table 2).

The angiocardiographic findings as illustrated in Fig. 4A, 4B, and 4C (Case 5) are considered to be specific for the diagnosis of this malformation. Other malformations in which a right-sided anatomical right atrium joins a right-sided anatomical left ventricle have to be distinguished angiocardiographically. In single left ventricle and l-(corrected) transposition of the great arteries, the right ventricle is reduced to an outflow chamber and, furthermore, in the great majority of the cases there exists transposition of the great arteries. In l-(corrected) transposition of the great arteries it is the pulmonary artery (instead of the aorta) which arises from the right-sided anatomical left ventricle, the aorta originating from the right ventricle, anterior and to the left of the pulmonary artery. The differential

diagnosis from mirror-image dextrocardia is less difficult; the aorta is posterior but situated to the left of the pulmonary artery, and there is a situs inversus of the viscera and atria.

Anatomically corrected transposition of the great arteries with l-loop should also be considered as a difficult angiocardiographic differential diagnosis. The presence of conal free wall (not only conal septum) below the aortic valve and above the right-sided morphological left ventricle should indicate anatomically corrected transposition of the great arteries with an l-loop. Below the two adjacent halves of both coronary aortic cusps of Case 1 (Case 5 of Tables 1, 2, and 3) there was a hypertrophied and misplaced conal septum, but there was no conal free wall to prevent the mitral-aortic fibrous continuity, below the remainder of the aortic valve. Thus, in this case there was no differential diagnostic problem from anatomically corrected transposition, for mitral-aortic fibrous continuity was demonstrated both angiographically (Fig. 4A and 4C) and anatomically (Fig. 5).

The basic physiological disturbance in isolated

<i>Tricuspid valve</i>	<i>Mitral valve</i>	<i>Morphological right ventricle</i>	<i>Ventricular septal defect</i>	<i>Conal septum</i>	<i>Atrial septum</i>	<i>Other</i>
Smaller than mitral valve Smaller than mitral valve	Normal	Small Smaller than morphological left ventricular; bioventricular hypertrophy	One slit-like defect 2 defects; one subaortic (membranous), one muscular	Normal	Patent foramen ovale Sealed foramen ovale	Coarctation of aorta Coarctation; persistent ductus arteriosus; aortic mitral continuity
Enlarged malformed with swellings in leaflets Normal	Normal Normal	Normal Large	One behind conal septum	Prominent	Sealed foramen ovale Sealed foramen ovale	Persistent ductus arteriosus Persistent ductus arteriosus; aortic mitral continuity
Hypoplastic supraavalvular fibrous ring (see dimensions in text)	Normal	Slightly small (see text for dimensions)	2 defects (see text)	Excessively developed and displaced towards morphological left ventricle	Sealed foramen ovale	Persistent ductus arteriosus; aortic mitral continuity
Hypoplastic supraavalvular fibrous ring (see dimensions in text)	Normal	Small (see text for dimensions)	One subaortic	Over morphological right ventricle; displaced as in Fallot's tetralogy	Sealed foramen ovale	Persistent ductus arteriosus; aortic mitral continuity

ventricular inversion (Fig. 1) is similar to that of complete transposition of the great arteries. Similar types of diagnostic (including the catheterization of the pulmonary artery) and surgical (balloon atrioseptostomy, Blalock-Hanlon operation, pulmonary artery banding, Mustard operation) procedures should, therefore, be indicated in patients with these malformations. There are certain anatomical features in isolated ventricular inversion which make management more difficult, both diagnostically and surgically. A sealed foramen ovale, constantly present in all previously reported cases of isolated ventricular inversion, made balloon atrial septostomy impossible and catheterization of the

FIG. 7 *Anatomical findings. Case 2 (Case 6 in Tables 1, 2, and 3). Internal view of the right-sided left ventricle. Observe the fibrous continuity (*) between the aortic (Ao) and mitral (M) valves. Note the smooth appearance of the left ventricular aspect of the ventricular septum (VS). The limits of the ventricular septal defect (VSD) are muscular.*

FIG. 8 Case 2 (Case 6 in Tables 1, 2, and 3). Internal view of the left-sided right ventricle. Observe the pulmonary valve (P) separated from the tricuspid valve (T) by the crista supraventricularis (CS), which, as in Fallot's tetralogy, is misplaced anteriorly to the septal band. The ventricular septal defect (VSD) is consequently located between the anterior (a) and posterior (p) divisions of the septal band (SB).

pulmonary artery more difficult. Only in Case 5 was there a ventricular septal defect related to the pulmonary artery, permitting this vessel to be catheterized from the right side of the heart.

Although the Mustard procedure to correct the malformation would theoretically be indicated for these patients, the high incidence of small-sized morphological right ventricles (in Cases 1, 5, and 6) and left atrioventricular valve obstructions (all cases except Case 4) would certainly limit its successful application in them.

The misplaced conal septum in Case 5 was also very hypertrophied (Fig. 4A, 5, and 6), a feature that has been encountered in some other specimens of our necropsy material in which there was a misalignment between the conal and ventricular septa.

As in other malformations with a discordance between the bulboventricular loop and the direction of the apex (a d-loop with a cardiac apex pointing to the left), in which a rotation of the heart

around its longitudinal axis has brought the left ventricle forward, a side-by-side ventricular spatial relation existed. Angiocardiographically, therefore, complete separation of the ventricles is observed in the frontal projection (Fig. 4A and 4B); in the lateral view these chambers are superimposed (Fig. 4C).

In this malformation, there is an inversion of the great arteries relative to the bulboventricular loop, a characteristic that has also been found in some of our necropsy cases of dextroversio cordis and anatomically corrected transposition of the great arteries; this is probably responsible for the discordance between the bulboventricular loop and the position of the great arteries encountered in all these situations (Quero Jiménez *et al.*, 1973; R. Van Praagh, 1973, personal communication).

Other important malformations in these cases of isolated ventricular inversion were: persistent ductus arteriosus in Cases 2, 3, 4, 5, and 6, and aortic coarctation in Cases 1 and 2 (Table 3).

A pulmonary artery banding and Blalock-Hanlon operation were performed in Cases 2 and 5, respectively. Both died soon afterwards of heart failure. The remaining patients died without surgery.

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