An Unusual Form of the Transposition Complex

Uncorrected Levo-transposition with Horizontal Ventricular Septum: Report of Two Cases


SUMMARY A distinctive angiographic appearance is described in two patients who had uncorrected levo-transposition of the great vessels. Although levo-transposition with inversion of the ventricles usually results in physiologically corrected transposition, in these patients the anatomy was arranged in such a way as to result in an uncorrected transposition. The following elements were responsible for this physiological condition: normal atrial situs, inverted ventricles with "criss-cross" atrioventricular flow, levo-transposed great arteries. In addition, the morphological right ventricle was hypoplastic, left sided and superior to the left ventricle and the ventricular septum was horizontal in position. The two ventricles were connected via a large ventricular septal defect. The importance of accurate, detailed, preoperative angiographic demonstration of the anatomic situation is stressed.

Case 1

A 14-year-old girl with cyanotic congenital heart disease was admitted to Stanford University Hospital in August 1974. A heart murmur was initially detected at age 3 months. Although moderately cyanotic she had no cyanotic spells, had normal developmental milestones, and did relatively well with near normal exercise tolerance throughout childhood. Cardiac catheterization was performed at age 7 years and demonstrated transposition of the great vessels. In the few months prior to the current catheterization, the patient noted increasing fatigue and a reduction in exercise tolerance.

Physical examination revealed a slender girl with moderate cyanosis and digital clubbing. The cardiac impulse was palpably increased. Auscultation revealed a loud pulmonic second sound, a grade 3/6 ejection systolic murmur along the left sternal border, a grade 2/6 diastolic murmur at the apex and an ejection click. The remainder of the physical examination was unremarkable. Electrocardiogram showed right axis deviation, biventricular hypertrophy, and probable bialtrial hypertrophy. A chest roentgenogram showed cardiomegaly and enlarged central pulmonary arteries. The pulmonary vasculature was markedly increased on the right and mild to moderately increased on the left. The hemoglobin level was 14.9 g% and the hematocrit 44.9%.

Cardiac catheterization demonstrated systemic pressures in both ventricles with identical pressures in the pulmonary artery and aorta and no gradient across either the pulmonary or aortic valves. Total pulmonary flow was calculated at 14.2 L/min/m²; the systemic flow was 3.7 L/min/m². There was a left-to-right shunt of 11.8 L/min and a right-to-left shunt of 1.3 L/min. Pulmonary vascular resistance was 3.3 units with a pulmonary to systemic vascular resistance ratio of 1:6.

Selective angiograms were performed in the right atrium (fig. 1), right ventricle (fig. 2), and left ventricle (fig. 3). The catheter was passed via the inferior vena cava into the right atrium which was situated in a solitus position, and then into a small anatomic right ventricle located to the left...
and above the anatomic left ventricle. The right atrial injection showed the right atrium to be in continuity with the inflow portion of the right ventricle; the systemic venous flow was from right atrium to right ventricle through a small right atrioventricular valve (fig. 1). The catheter was advanced through the small tricuspid valve into the anatomic right ventricle which consisted of small sinus and infundibular portions (fig. 2). The aorta arose from the right ventricle. It was small and situated anteriorly and to the left of the main pulmonary artery (figs. 2, 3, right).

The catheter was then introduced, via a large ventricular septal defect, into a large anatomic left ventricle lying inferiorly (fig. 3). The two ventricles were separated by a septum positioned in the horizontal plane (fig. 3, right). Bidirectional flow through the ventricular septal defect correspondingly was in a vertical direction between the inferiorly positioned left ventricle and the superiorly placed right ventricle. The left atrioventricular valve was demonstrated on the left ventricular injection (fig. 3, right). Continuity between left atrium and anatomic left ventricle was established during the levophase when contrast sequentially opacified the left atrium and anatomic left ventricle. The pulmonary artery arose from the left ventricle and the pulmonic valve was at a lower level than the aortic valve. The main pulmonary artery and central pulmonary arteries were markedly dilated and tortuous but there was normal gradual tapering of peripheral pulmonary arteries. The coronary arterial pattern was characteristic of inversion of the ventricles with the left anterior descending branch arising from the right coronary artery (fig. 4, left). In addition the course of the left anterior descending branch was unusual; like the interventricular septum, it was oriented horizontally.

Case 2

An 11-month-old boy was admitted to Stanford University Hospital in October 1974 with cyanotic congenital heart disease and marked growth retardation. A cardiac murmur and cyanosis were detected on the first day of life. Because of severe hypoxemia he underwent a cardiac catheterization which demonstrated transposition of the great vessels. A balloon atrial septostomy was performed. Because of persistent severe hypoxemia, a Blalock-Hanlon atrial septostomy was performed and resulted in improvement in his clinical status.

Physical examination revealed a cyanotic child with height and weight below the third percentile. Palpation revealed a right ventricular impulse and a faint systolic thrill along the left sternal edge. The first heart sound was normal, the second single and there was a grade 4/6 long ejection systolic murmur widely transmitted but maximal at the left sternal edge. Apart from a pectus excavatum and a thoracotomy scar, the remainder of the physical examination was unremarkable. The electrocardiogram revealed sinus rhythm with periods of junctional rhythm and atrioventricular dissociation. There was right axis deviation.

**FIGURE 1** Anteroposterior view of right atrial injection (Case 1) showing right atrium in situ solitus. Contrast opacifies right atrium in continuity with right ventricular inflow tract. Reflux contrast opacifies the inferior vena cava and hepatic veins. Note faint opacification of aorta and absence of opacification of the left ventricle in this radiograph obtained soon after injection. A = aorta; H = hepatic veins; RA = right atrium; RV = right ventricle; t = tricuspid valve; VC = inferior vena cava.

**FIGURE 2** Lateral view of right ventriculogram (Case 1). Small aorta arises from infundibular portion of anterior rudimentary right ventricle and lies anterior to large pulmonary artery. A = aorta; LV = left ventricle; P = main pulmonary artery; RV = right ventricle.
Figure 3  Frontal view of the left ventriculogram. Left) The catheter enters the large left ventricle via a ventricular septal defect outlined between the arrows. Right) At a later phase, the superior right ventricle is filled via the ventricular septal defect and the aorta and pulmonary trunk opacified. The aorta is positioned to the left of the pulmonary trunk (levo-transposition). The horizontally positioned ventricular septum between the superior small right ventricle and the larger, inferior left ventricle is well demonstrated. A = aorta; LV = left ventricle; m = mitral valve; P = pulmonary trunk; RV = right ventricle.

(+150), right ventricular hypertrophy and biatrial hypertrophy. Q waves were present in the right precordial leads and absent in the left precordial leads. Chest X-ray revealed normal cardiac size and a prominent left superior cardio-mediastinal border representing the levo-transposed aorta. Pulmonary vascularity was considered normal. The hemoglobin level was 17.2 g %, and the hematocrit 49.5%.

Cardiac catheterization demonstrated systemic pressures in both ventricles and the aorta. Total pulmonary flow was calculated at 3.9 L/min/m² and systemic flow at 6.4

Figure 4  The coronary arterial pattern in both Case 1 (left) and Case 2 (right) is characteristic for ventricular inversion. The right coronary artery which now encircles the anatomic left ventricle has the anatomic distribution of a normal left coronary artery. It gives origin to the anterior descending coronary artery which follows a course corresponding to the orientation of the ventricular septum. A = aorta; RV = right ventricle; → = right coronary artery; " " = anterior descending artery.
L/min/m². The pulmonary-to-systemic flow ratio was 0.6:1.0. A left-to-right shunt was calculated at 0.7 L/min and there was a right-to-left shunt of 1.5 L/min. Pulmonary vascular resistance was 3.8 units and systemic vascular resistance 25.7 units. Selective angiocardiograms were performed in the right ventricle (fig. 5), left ventricle (fig. 6) and the ascending aorta (fig. 4, right). The small anatomic right ventricle was oriented horizontally, above, anterior and to the left of the anatomic left ventricle (fig. 5). Tricuspid regurgitation during the right ventricular injection showed that the right ventricle was in continuity with the right atrium, which was in the solitut position (fig. 5). The aorta, situated anterior and to the left of the pulmonary artery, arose from the anatomic right ventricle (figs. 5, 6); the aortic valve was located above the level of the pulmonary valve. Left ventricular injection demonstrated a normal sized anatomical left ventricle lying inferior and to the right of the ventricular septum which was nearly horizontal in position (fig. 6). A large ventricular septal defect was present. There was a double conus (figs. 5, left, 6, right) with discontinuity between the two atroventricular valves and both of the semilunar valves. During the levophase, continuity between left atrium and left ventricle was established and blood flow was from left atrium to left ventricle. The infundibulum below the pulmonary valve was short and narrow and there was severe valvar pulmonary stenosis (fig. 6, right). The coronary arterial anatomy was the characteristic type associated with ventricular inversion; the left anterior descending coronary artery arose from the right coronary artery and tended to follow a horizontal course, conforming to the peculiar position of the ventricular septum (fig. 4, right).

**Discussion**

The current cases are examples of an unusual variant of the transposition complex. The anatomic relationships in each case consisted of levo-transposition of the great vessels and inversion of the ventricles. However, since the inverted right ventricle maintained connection with the right atrium and the left ventricle maintained its connection to the left atrium, the pulmonary and systemic circulations were in parallel and thus the transpositions were physiologically uncorrected. This association of inversion of the ventricles with a physiologically uncorrected transposition is very rare in-

**FIGURE 5** Sequential (opposite; below, left and right) anterior-posterior frames from right ventriculogram of Case 2. The right atrium was in direct continuity with the right ventricle via the inflow portion of the right ventricle. The aorta arises from the infundibular portion of the right ventricle and lies in the levo-position. Note the diminutive but distinct sinus and infundibular portions of the inverted right ventricle. $A = $ aorta; $i = $ infundibular portion of right ventricle; $LV = $ left ventricle; $P = $ pulmonary artery; $RA = $ right atrium; $RV = $ right ventricle; $s = $ sinus portion of right ventricle; $t = $ tricuspid valve; $→$ $ =$ subpulmonary conus; $→→$ = outlining ventricular septal defect.
To the best of our knowledge the angiographic demonstration of the anatomy of such a case has not been previously described in detail.

Angiography demonstrated levo-transposed great vessels with the aorta arising from a small morphologic right ventricle. The ventricular septum was oriented in almost a horizontal plane and thus the right ventricle tended to be positioned superiorly and the larger left ventricle inferiorly. The position of the great vessels and ventricles in these two cases is identical to a single case reported by Kinsley et al.6 However, this latter case was physiologically corrected since the left atrium was in solitus position and the right ventricle by virtue of a small tricuspid valve. Similarly, the left atrium occupied a normal position and was anatomically in continuity with the right ventricle via a small tricuspid valve. These relationships of the atria and ventricles resulted in apparent crossing of the inflow portions of each ventricle in the frontal angiocardiogram (fig. 7). This type of relationship has previously been termed "criss-cross ventricles." The description of the anatomic specimen in one of the cases reported by Anderson et al.6 bears a close resemblance to the current cases. Similarly, Van Praagh7 has described a heart with transposition of the great vessels in which the atria were in solitus position and the ventricles inverted; as in the current cases, the left-sided anatomic right ventricle received right atrial blood.

In both of the present cases the right ventricles were diminutive, but distinct sinus and infundibular portions were demonstrated. There are similarities in the angiocardiographic appearance of the present cases with several cases recently described of common ventricle with a subaortic infundibular chamber.8 However, the two cases we have described may be separated from the spectrum of common ventricle by the presence of distinct, although diminutive inflow and infundibular portions in the right ventricle (fig. 5).

Case 1, by virtue of a large ventricular septal defect which permitted optimal intracardiac mixing of the systemic and pulmonary circulations, remained minimally symptomatic into adolescence. On the other hand, Case 2 possessed a similar type of ventricular septal defect but was profoundly hypoxemic in the immediate neonatal period. This was a consequence of severe subvalvular and valvar pulmonic stenosis.

The coronary arterial pattern in the current cases was that
characteristic of ventricular inversion. In each case the anterior descending coronary artery arose from the proximal right coronary artery; i.e., the right coronary had the anatomic configuration of a left coronary artery. Moreover, the anterior descending and posterior descending branches ran in a roughly horizontal rather than vertical fashion; this course corresponded to the horizontally oriented ventricular septum.

Precise definition of the intracardiac and coronary arterial anatomy is essential in considering the feasibility of and planning the method for total correction of these complicated defects. The major limiting factor to successful total correction, particularly in Case 2, is the small size of the anatomic right ventricle.

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