

Isolated Ventricular Inversion and Anatomically Corrected Malposition of the Great Arteries Associated with Right Juxtaposition of Left Atrial Appendage

—A case of successful surgical repair—

—Abstract—

Jeong Ryul Lee, M.D., Yong Jin Kim, M.D.,
Kyung Phill Suh, M.D., and Yong Soo Yoon, M.D.**

A seven month old female infant with isolated ventricular inversion and anatomically corrected malposition of the great arteries in situs solitus, associated with ventricular septal defect, patent ductus arteriosus, right-sided juxtaposition of left atrial appendage, is reported. The patient showed usual atrial arrangement with somewhat superoinferior relation, a discordant atrioventricular connection, and a concordant ventriculoarterial connection with aorta in the right-sided position. A normal sized left atrium was connected to the left superiorly positioned morphologic right ventricle through a tricuspid valve, which crossed the left ventricular outflow tract anteriorly. Well developed bilateral(subaortic and subpulmonary)conus was documented at operative field. successful surgical repair was done by performing the Senning procedure and by closing the ventricular septal defect with a patch through the right ventriculotomy. The infant's postoperative course was uneventful with normal sinus rhythm. Postoperative cardiac catheterization revealed no hemodynamic obstruction or residual shunt.

Introduction

Inversion of the ventricle without transposition of the great arteries with situs solitus is rare congenital heart anomaly, previously having been reported eight surgically corrected cases until 1985¹⁾, also those combined with anatomically corrected malposition of the great arteries as ours is still further rare^{10,11,12)}. The cyanosis is the main clinical

symptom and blood flow in this anomaly is analogous to that found in complete transposition of the great arteries. Right-sided Juxtaposition of left atrial appendage, which is shown to be associated with a discordant atrioventricular connection and atresia of the tricuspid valve with significant frequency, is eight times less common than left juxtaposition²⁾, and at least twenty five autopsied cases have been reported³⁾.

We report our experience with this rare congenital cardiac anomaly in seven months old female infant and provide the review of the literature.

*Department of Thoracic and Cardiovascular Surgery,
College of Medicine, Seoul National University

**Department of Pediatrics, College of Medicine,
Seoul National University.

1990년 10월 24일 접수

Case Report

A seven month old female infant was noticed to have sweating and dyspnea with deepening cyanosis since birth. Her gestational age was 40 weeks and the birth weight was 3.15 kg. A subxyphoid impulse was present with precordial bulging on external inspection. Physical examination revealed accentuated second heart sound without any audible murmur. The peripheral pulse was normal and the liver was palpable one finger breadth from the right costal margin. Chest Roentgenogram showed a left cardiac apex, cardiomegaly, and increased pulmonary vascular markings (Fig. 1). The electrocardiogram was normal sinus rhythm with P wave and QRS axis 30 and 90 degrees. Echocardiogram revealed, situs solitus, atrioventricular discordant connection, rightsided aorta with subaortic conus, anteriorly positioned left sided atrioventricular valve crossing the outflow tract of right sided morphologic left ventricle with large subaortic ventricular septal defect (Fig. 3A, B). At cardiac catheterization, mean pulmonary arterial pressure was 52 mmHg; pulmonary vascular

resistance 7.2u; both ventricular systolic pressure 70 mmHg with higher oxygen saturation of the left sided ventricle. Angiocardiogram revealed a right sided inferior vena cava and a left sided stomach bubble; the morphology of the right sided and posterior ventricle was typically left ventricle; the left-sided ventricle was morphologically right ventricle with its atrioventricular valve crossing the outflow tract of right sided ventricle; the aorta arising at the right side of pulmonary artery,

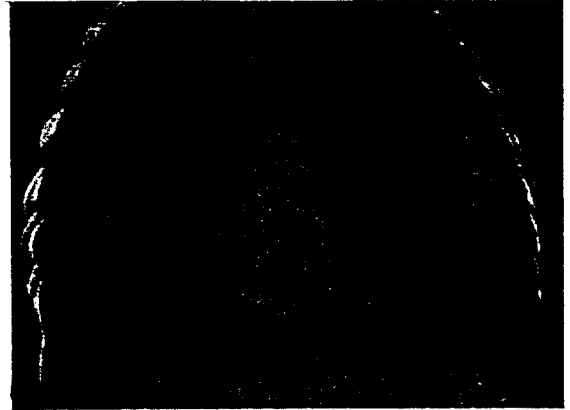


Fig. 1. Admission chest roentgenogram showed cardiomegaly with increased pulmonary vascularity.

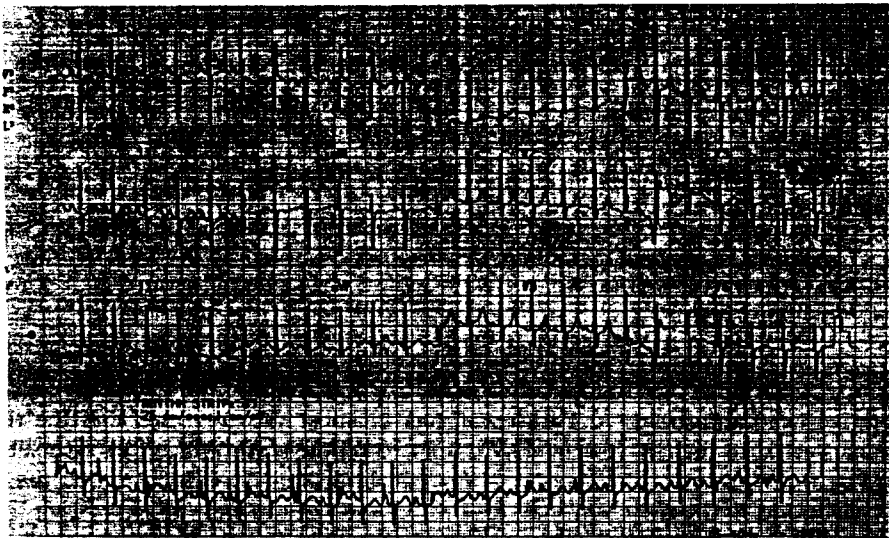


Fig. 2. Preoperative electrocardiogram was normal sinus rhythm with biventricular hypertrophy.

from the morphologic left ventricle; the pulmonary artery originating from the morphologic right ventricle: Both atrial appendages were right-sided juxtaposed, : A large subaortic ventricular septal defect was present (Fig. 4A~D).

At the age of seven months, the infant became more cyanotic and corrective operation was performed on Aug 23th 1990. Grossly, the aorta was right to the pulmonary artery and both atrial appendages were right juxtaposed. Intracardiac observation through a right atriotomy revealed an intact atrial septum with a small patent foramen ovale and the right atrium connected to the morphologic left ventricle through a bicuspid mitral valve. Atrial septotomy revealed the left atrium connected to the morphologic right ventricle through tricuspid valve, which crossed the left ventricular outflow tract (Fig. 5A~C, Fig. 6). An intraatrial switch of venous return using the Senning procedure was performed only with the patient's own cardiac tissue. Ventricular septal defect was closed with a patch through a right ventriculotomy which revealed well developed subpulmonary conus. Rewarming and weaning from extracorporeal cir-

ulation was uneventful with the infant's central venous pressure of 18 cmH₂O, and left atrial pressure of 14 cmH₂O. He suffered from prolonged pleural effusion until the 16th postoperative day. Cardiac catheterization and angiocardiogram was performed one month after operation, which revealed no hemodynamic obstruction (Fig. 7).

Anatomical description: The arrangement of the abdominal organs and the lobation of lungs were normal, the heart was left sided with its apex pointing to the left. Atrial morphology and the relation between two atrial chambers were normal with right atrium located a little anteroinferior to the left atrium. Systemic and pulmonary venous connections were normal. The atrioventricular connection was discordant with two separate atrioventricular valves, the left of which was smaller than the right. The right atrium was connected to right sided morphologic left ventricle through a bicuspid mitral valve. The subvalvular apparatus was normal. The morphologic left ventricle was connected to a right posterior aorta. The anterior leaflet of the mitral valve was separated from the aortic cusp by subaortic conus around 1cm, the

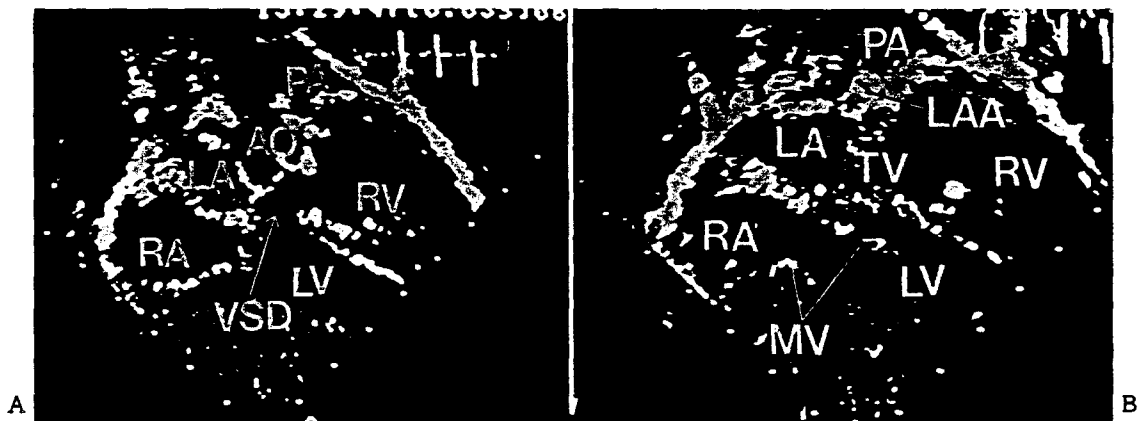


Fig. 3. Preoperative echocardiogram : A, Parasternal four chamber view revealed that the right atrium (RA) is connected to the left ventricle (LV), from which the aorta (AO) arises with subaortic conus. B, More frontal plane of the parasternal four chamber projection discloses that the left atrium (LA) is connected to the right ventricle (RV) with its atrioventricular valve crossing the left ventricular outflow tract anteriorly. The ventricle is connected to the pulmonary artery. Left atrial appendage (LAA) is in front of aortic pole VSD=ventricular septal defect; MV=mitral valve; TV=tricuspid valve.

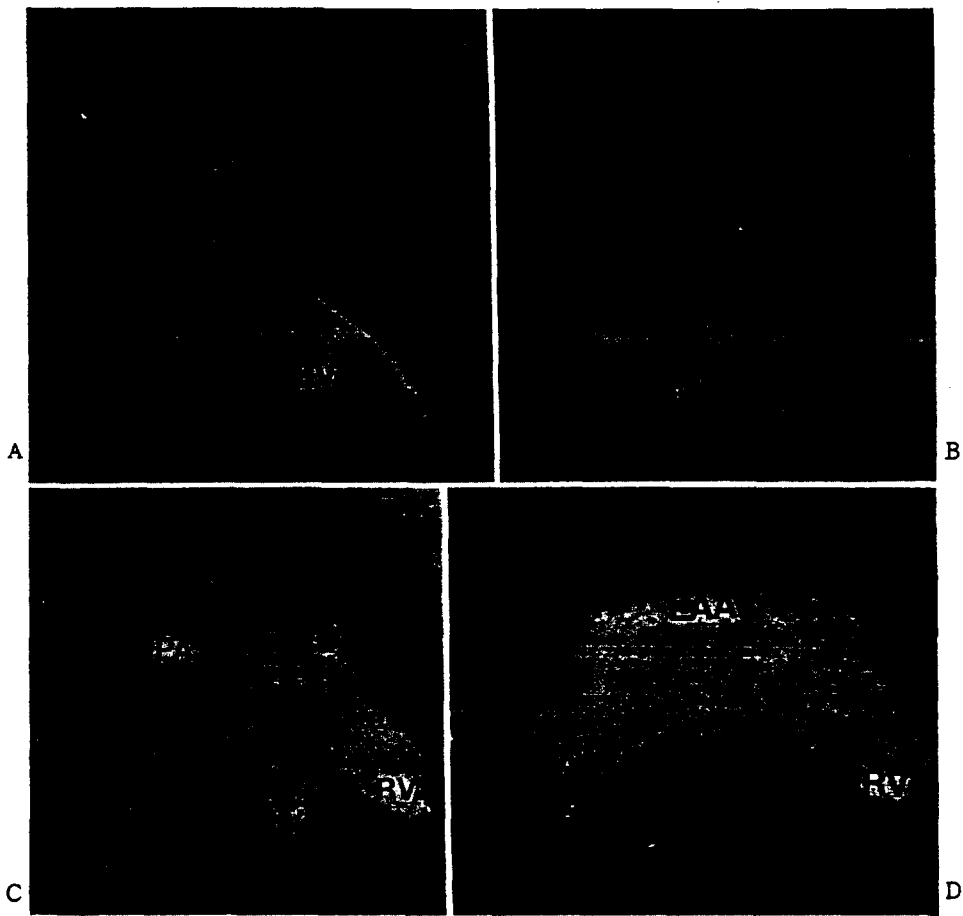


Fig. 4. Preoperative angiocardigram : A, Anteroposterior projection of left ventriculogram ; The left ventricle(LV) ejects through a subaortic conus(SAC) into the aorta(AO), which is to the right of the pulmonary artery(PA). B, Lateral film of left ventriculogram shows subaortic conus and dye leakage through subaortic ventricular septal defect(VSD). C, Anteroposterior projection of right ventriculogram at systolic phase : The right ventricle(RV) ejects into the left sided pulmonary artery. D, Anteroposterior projection of right ventriculogram at diastolic phase ; Left atrium ejects into the left sided morphologic right ventricle, with atrioventricular valve crossing the left ventricular outflow tract anteriorly. Left atrial appendage shows right-sided juxtaposition. AA=ascending aorta.

ventricular septum was abnormally oriented, so that its right inferior aspect faces the right sided morphologic left ventricle and the left superior aspect does the left sided morphologic right ventricle. A perimembranous large ventricular septal defect was present. The left atrium was connected to the left superiorly located morphologic right ventricle through the tricuspid valve, which was smaller than the right sided mitral valve in diameter. Left atrial appendage was right-

side juxtaposed to the right atrial appendage and was anterior to the aortic pole. The normal sized ventricle had a well developed crista supraventricularis and was connected to the left-sided pulmonary artery with an infundibulum.

Comments

Van Praagh and Van Praagh⁴⁾ uses the term isolated ventricular inversion in 1966, while Shi-

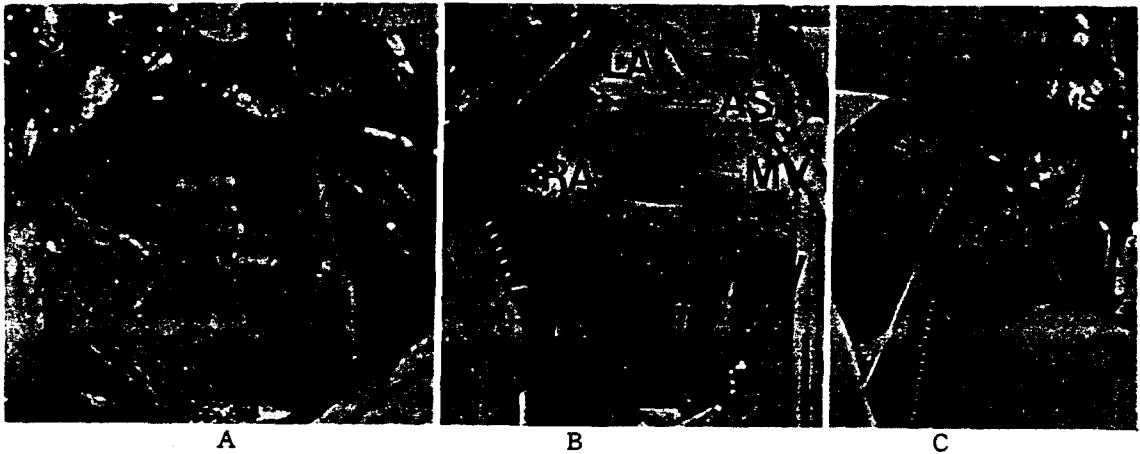


Fig. 5. Operative findings : A, External cardiac morphology : Superiorly located left atrium (LA) with its right juxtaposed auricle (LAA) is connected to the left superiorly located morphologic right ventricle (RV), from which pulmonary artery (PA) arises at the left side of aorta. Inferiorly located right atrium (RA) is connected to the right inferiorly positioned morphologic left ventricle (LV), from which the aorta (AO) arises at the right side of pulmonary artery. B, Both atriotomy shows somewhat horizontally positioned atrial septum (AS). Superior atrioventricular valve has a tricuspid valve (TV) morphology and the inferior one does a bicuspid mitral valve (MV). C, Right ventriculotomy discloses the moderate sized ventricular septal defect (VSD) with subpulmonary conus (SPC).

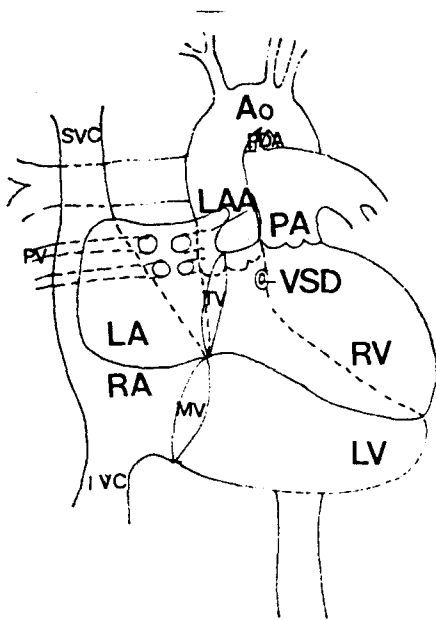


Fig. 6. Schematic drawing summarizing the cardiovascular malformation : Left atrium (LA) crosses the left ventricular outflow tract anteriorly with its auricle (LAA) right side juxtaposed. Right atrium (RA) is connected to the morphologic left side juxtaposed. Right atrium (RA) is connected to the morphologic left ventricle (LV) through a morphologic mitral valve (MV), while left atrium is connected to the morphologic right ventricle (RV) through a morphologic tricuspid valve (TV). The ventriculoarterial relation is concordant. Subaortic ventricular septal defect (VSD) is present and the conus is bilateral. A=aorta : PA=pulmonary artery : SVC=superior vena cava : IVC=inferior vena cava : PV=pulmonary vein.

neborne et al.⁵⁾ described this anomaly as visceratrial situs solitus, discordant atrioventricular and concordant ventriculoarterial connection, using segmental approach. Calbro and colleagues⁶⁾

pointed out that isolated atrioventricular discordance requires the presence of a well-defined situs and of both ventricles regardless of their spatial relation, and to describe and define this

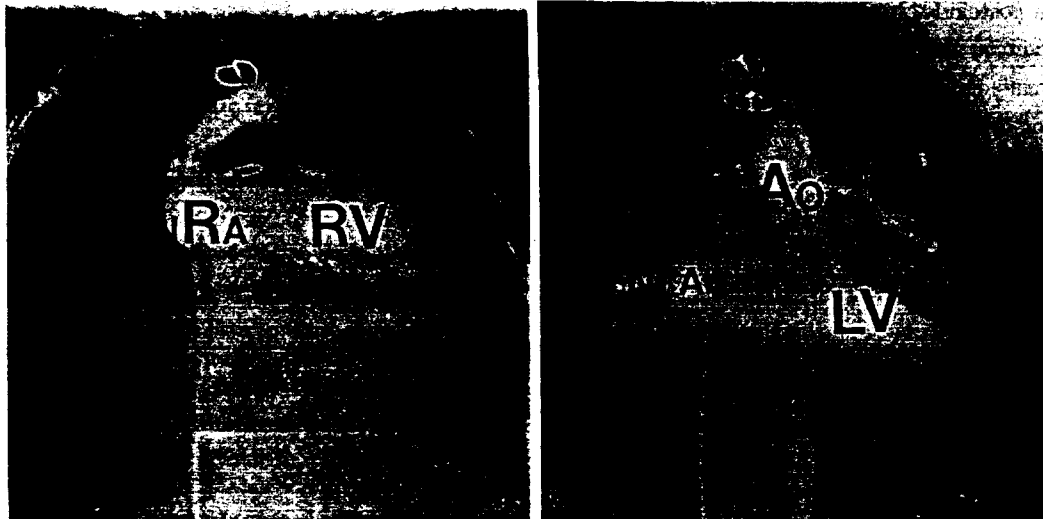


Fig. 7. Angiocardiogram one month after Senning operation, which revealed no hemodynamic obstruction or anatomic narrowing. LV=left ventricle : RV=right ventricle : Ac=Aorta : NRA=neo-right atrium : NLA=neo-left atrium

anomaly correctly, intracardiac circulation should be stressed first, and then the relation between the chambers. In this anomaly, the ventriculo-arterial connection must be concordant, with normally related great arteries. The most frequently occurring infundibular morphology is represented by mitral aortic continuity and muscular discontinuity between the tricuspid valve and the pulmonary artery. Ventricular septal defect^{1,3,7)}, aortic stenosis⁴⁾, patent ductus arteriosus^{3,7)}, complete atrioventricular canal⁸⁾, left atrioventricular valve atresia^{3,9)} have been reported as frequent associated anomalies.

A terminology of "anatomically corrected malposition of the great arteries (ACM)" is defined variably. Anderson et al¹⁰⁾, described this anomaly as that in which the great arteries arise in unusual fashion from their morphologically appropriate ventricles, emphasizing that ACM describes not a discrete anomaly but only a ventriculo-arterial relation, which is one of ventriculoarterial concordance, and furthermore can coexist with all varieties of atrioventricular relations. Van Praagh and colleagues¹¹⁾ established the fact that ACM can occur with a subaortic conus, as well as with a

bilateral conus as ours. Kirkin et al¹²⁾ reported successful surgical repair in 2 patients with ACM, but their cases had atrioventricular concordance unlike our case.

Collective reviews and discussion of the morphologic characteristics of hearts with right sided juxtaposition of the atrial appendages¹²⁻¹⁸⁾ have been made by several authors. Significance of right juxtaposition of atrial appendages in relation to the association with severe cyanotic heart disease is stressed by Melhuish and Van Praagh in 1984¹⁴⁾, but Anderson et al¹⁹⁾ contended that it does not always associate with severe complex cardiac lesions, and even it could be present as an isolated lesion. Nonetheless, frequent association with discordant atrioventricular connection and atresia of tricuspid valve were reported at several articles^{3, 13)}. Seo et al. collected 25 autopsied cases of an unusual ventricular loop associated with right juxtaposition of the atrial appendages suggesting that the embryologic mechanism producing disharmony between the atrioventricular connection and the segmental combinations be interpreted on the basis of posterior ventricular looping, since they are best explained on the basis of a hypothetical heart

with posteriorly located outflow tract. Our case was similar to Seo's case 2 except the presence of normal left sided atrioventricular valve (morphologic tricuspid valve), which is anterior to the aortic pole^{3,20}.

From the clinical standpoint, cyanosis is most common symptom and vectocardiogram shows early left anterior vectorial forces which is thought to be the result of the abnormal position of ventricular septum⁶. Pasquini and colleagues²¹ contended that multiple plane two-dimensional echocardiography can provide excellent segmental analysis including visceratrial situs, ventricular loop and great arterial position and a surgical approach through the left sided infundulum affords excellent exposure of the ventricular septal defect and atrioventricular conduction block^{8,22}, which occurs frequently as a postoperative complication, can be avoided. Nonetheless, the final diagnosis of this anomaly should be done by cardiac catheterization and angiocardiographic data. The definitive surgical management of this cyanotic malformation consists of redirection of venous blood flow toward their respective arteries by performing an intraatrial venous switch using Senning^{1,24} or Mustard procedures^{8,22,23}. The literatures about surgically corrected cases of isolated ventricular inversion were reviewed by Baudet et al¹¹. with their case, which suggested the mortality rate was higher, especially for palliative procedures, when additional defects were present. Of eight cases, two patients died early post operatively of low cardiac output, two have had a complete heart block postoperatively. Our case showed normal sinus rhythm with right bundle branch block postoperatively.

REFERENCES

1. Baudet EM, Hafez A, Choussat A, Roquez X : *Isolated ventricular inversion with situs solitus ; Successful surgical reapir. Ann Thorac Surg 41:91,*

1986

2. Boor PJ, McAllister PR : *Juxtaposition of the atrial appendages. Hum Pathol 10: 347, 1979*
3. Seo JW, Choe JY, Chi JG : *An unusual ventricular loop associated with right juxtaposition of the atrial appendages. Int J Cardiol 25: 219, 1989*
4. Van Praagh R, Van Praagh S : *Isolated ventricular inversion : a consideration of the morphogenesis, definition and diagnosis of non transposed and transposed great arteries. Am J Cardiol 17 : 395, 1966*
5. Shinebourne EA, Macartney FJ, Anderson RH : *Sequential chamber localization : logical approach to diagnosis in congenital heart disease. Br Heart J 38 : 327, 1976*
6. Calabro R, Marino B, Marcico F : *A case of isolated atrioventricular discordance. Br Heart J 47 : 400, 1982*
7. Jimenez M Q, Raposo-Sonnenfeld I : *Isolated ventricular inversion with situs solitus. Br Heart J 37 : 293, 1975*
8. Hazan E, Baillet F, Dupuis C : *Isolated ventricular discordance and complete atrioventricular canal in situs inversus : report of successful surgical repair. Am Heart J 99 : 163, 1977*
9. Dunkman WB, Perloff JK, Roberts WC : *Ventricular inversion without transposition of great arteries : A rarity found in association with atresia of the left sided(tricuspid) atrioventricular valve. Am J Cardiol 39 : 226, 1977*
10. Anderson RH, Becker AE, Losekoot TG, Gerlis LM : *Anatomically corrected malposition of the great arteries. Br Heart J 37 : 993, 1975*
11. Van Praagh R, Durnin RE, Jockin HJ, Wagner HR, Kornis M, Garabedian H, Ando M, Calder A : *Anatomically corrected malposition of the great arteries {S, D, L}. Circulation 51 : 20, 1975*
12. Kirklin JW, Pacifico AD, Bargeron, Jr. LM, Soto B : *Cardiac repair in anatomically corected malposition of the great arteries. Circulation XLV-III : 153, 1973*
13. Charuzi Y, Spanos PK, Amplatz K, Edwards JE : *Juxtaposition of the atrial appendages. Circulation XLV II : 620, 1973*
14. Melhuish BPP, Van Praagh R : *Juxtaposition of atrial appendages : a sign of severe cyanotic congenital heart diseae. Br Heart J 30 : 269, 1968*

15. Becker AE, Becker MJ : *Juxtaposition of atrial appendages associated with normally oriented ventricles and great arteries. Circulation XLI:685, 1970*
16. Wagner HR, Aldey LE, Vlad P : *Juxtaposition of atrial appendages : A report of six necropsied cases. Circulation XLII : 157, 1970*
17. Mathew R, Replogle R, Thilenius OG, Arcilla RA : *Right juxtaposition of atrial appendages. Chest 67 : 483, 1975*
18. Embree J, Aterman K, Belcourt C : *Right sided juxtaposition of the atrial appendages in an anencephalic. J Pathol 129 : 157, 1979*
19. Anderson RH, Smith A, Wilkinson JL : *Right Juxtaposition of the auricular appendages. Eur J Cardiol 4 : 495, 1976*
20. Anderson RH, Smith A, Wilkinson JL : *Dis-harmony between atrioventricular connections and segmental combinations : unusual variants of "crisscross" hearts. J Am Coll Cardiol 10 : 1274, 1987*
21. Pasquini L, Sanders SP, Parness I, Colan S, Keane JF, Mayer JE, Kratz C, Foran RB, Marino B, Van Praagh S, Van Praagh R : *Echocardiographic and anatomic findings in atrioventricular discordance with ventriculoarteral concordance. Am J Cardiol 62 : 1256, 1988*
22. De Leval MR, Bastos P, Stark J, et al : *Surgical technique to reduce the risks of heart block following closure of ventricular septal defect in atrioventricular discordance J Thorac Cardiovasc Surg 78 : 515, 1979*
23. Ostermeyer J, Birks W, Krian A, Sievers J, hilgenberg F : *Isolated atrioventricular discordance : report of two surgical cases with isolated ventricular inversion. J Thorac Cardiovasc Surg 86 : 926, 1983*
24. Fox LS, Kirklin JW, Pacifico RD, Waldo AL, Bargeron, Jr LM : *Intracardiac repair of cardiac malformations with atrioventricular discordance. Circulation 54 : 123, 1976*