Anatomically Corrected Malposition of the Great Arteries

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We report the case of an infant with anatomically corrected malposition of the great arteries in solitus {S, L, D}, associated with ventricular septal defect, patent ductus arteriosus, and right juxtaposition of the atrial appendages. The aortic pole was posterior to the tricuspid-mitral line and the conus was bilateral. Successful surgical repair was undertaken by the Senning procedure with closure of ventricular septal defect and division of the ductus. Postoperative cardiac angiography demonstrated no hemodynamic obstruction or residual shunt.

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Anatomically corrected malposition of the great arteries is a rare congenital cardiac anomaly in which the great arteries arise from their appropriate ventricles in an unusual spatial relationship [1–3]. Right juxtaposition of the atrial appendages has been shown to be significantly associated with a discordant atrioventricular connection [4]. This rare combination of congenital cardiac anomalies has been reported by several authors [4–6]. Cyanosis is the main clinical symptom, and blood flow in this anomaly is analogous to that found in complete transposition of the great arteries. We report our experience of successful surgical repair of this rare combination of congenital cardiac anomalies in a 7-month-old female infant.

A 7-month-old female infant was noticed to have dyspnea and cyanosis since birth. Her gestational age was 40 weeks, and the birth weight was 3.15 kg. On examination, she had an accentuated second heart sound without any audible murmur. Chest roentgenography showed a left cardiac apex. Echocardiography showed the left-sided atrioventricular valve crossing the outflow tract of the right-sided morphologic left ventricle. Angiocardiography demonstrated a right-sided inferior vena cava and a left-sided stomach bubble. The morphology of the right-sided ventricle was typically left ventricle, and a subaortic conus (aortomitral discontinuity) was seen (Figs 1A, B). The left-sided ventricle was morphologically the right ventricle, with its atrioventricular valve crossing the outflow tract of the right-sided morphologic left ventricle anteriorly (Fig 1C). The aorta arose to the right of and anterior to the pulmonary artery from the morphologic left ventricle (Fig 1B), whereas the pulmonary artery originated from the morphologic right ventricle. Right-sided juxtaposition of the atrial appendages (Fig 1C) and a large subaortic ventricular septal defect were present (Fig 1B).

Operation was done when the patient was 7 months old. Grossly, the aorta was right of and anterior to the pulmonary artery, and both atrial appendages were right juxtaposed. Intracardiac inspection 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 showed that the left atrium connected to the morphologic right ventricle through the tricuspid valve, which crossed the left ventricular outflow tract anteriorly. An intraatrial switch of venous return using the Senning procedure was performed. The ventricular septal defect was closed with a patch through a right ventriculotomy, which revealed well-developed subpulmonary conus. Rewarming and weaning from extracorporeal circulation were uneventful. The postoperative course was smooth except for prolonged pleural effusion until the 16th postoperative day. Cardiac catheterization and angiocardiography were performed 1 month after operation and disclosed no hemodynamic obstruction or residual shunt (Fig 2).

Comment

The term *anatomically corrected malposition of the great arteries* is defined variably. Anderson and associates [1] described this anomaly as the great arteries arising in unusual fashion from their morphologically appropriate ventricles, emphasizing that anatomically corrected malposition of the great arteries describes not a discrete anomaly but only a ventriculoarterial relation, which is one of ventriculoarterial concordant connection, and furthermore can coexist with all varieties of atrioventricular relations. Van Praagh and colleagues [2] included four possible varieties, depending on the segmental set or

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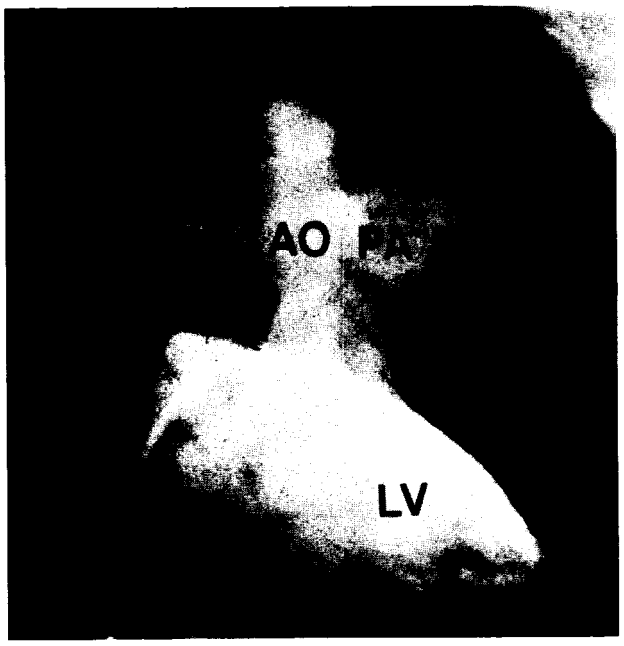
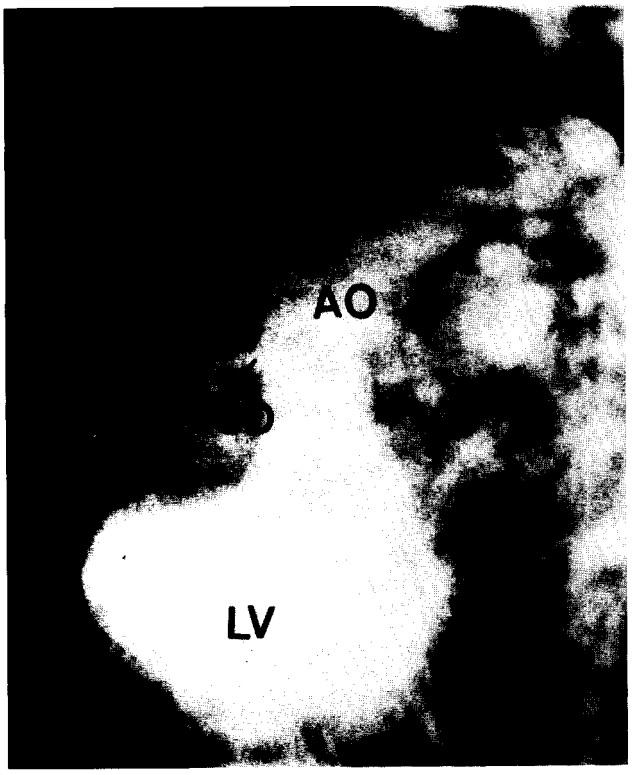


A



B

C

Fig 1. Preoperative angiocardiogram. (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 contrast medium leakage through subaortic ventricular septal defect* (VSD). *(C) Anteroposterior projection of right ventriculogram at diastolic phase: left atrium* (LA) *ejects into the left-sided morphologic right ventricle, with atrioventricular valve crossing the left ventricular outflow tract anteriorly. Left atrial appendage* (LAA) *shows right-sided juxtaposition.* (AA = *ascending aorta.*)

combination (ie, with atrioventricular concordant or atrioventricular discordant connections); they established the fact that anatomically corrected malposition of the great arteries can occur with a subaortic conus, as well as with a bilateral conus as in our patient. Kirklin and co-workers [3] reported successful surgical repair in 2 patients with anatomically corrected malposition of the great arteries, but their patients had atrioventricular concordant connections, unlike our patient.

The surgical significance of juxtaposition of the atrial appendages was reviewed by Anjos and associates [7], who emphasized that juxtaposition is almost always as-

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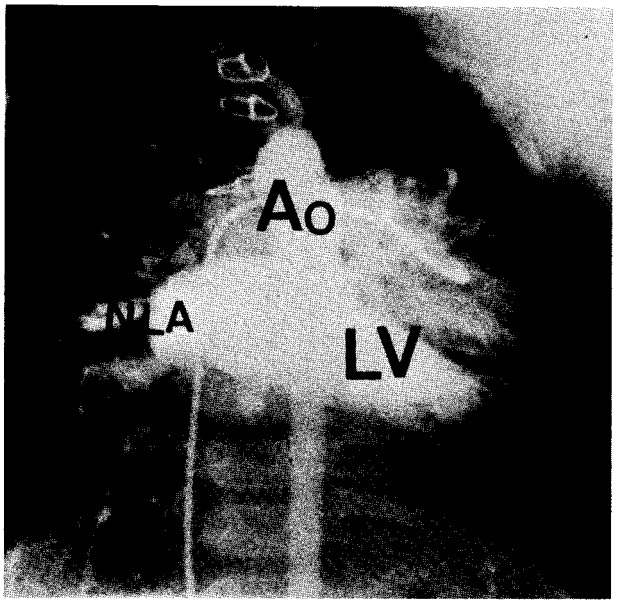

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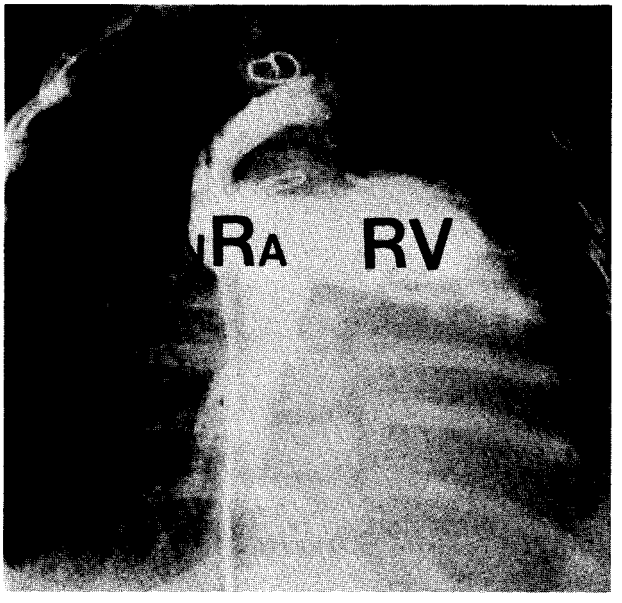

B

Fig 2. Postoperative angiocardiogram reveals no hemodynamic obstruction or anatomic narrowing. (Ao = *aorta;* LV = *left ventricle;* NLA = *neo-left atrium;* NRA = *neo-right atrium;* RV = *right ventricle.*)

sociated with anomalous segmental connections, particularly with a discordant ventriculoarterial connection and frequently with anomalous atrioventricular connections. But right juxtaposition is usually associated with less serious malformations, and it can even be present as an isolated lesion [8]. Seo and colleagues [4] collected 25 autopsied cases of an unusual ventricular loop associated with right juxtaposition of the atrial appendages and suggested that the embryologic mechanism producing disharmony between the atrioventricular connection and the segmental combinations be interpreted on the basis of posterior ventricular looping, as they are best explained on the basis of a hypothetical heart with a posteriorly located outflow tract. Our patient was similar to Seo and colleagues' patient 2 except for the presence of normal left-sided atrioventricular valve (morphologic tricuspid valve), which was anterior to the aortic pole.

From the clinical standpoint, cyanosis is the most common symptom. Final diagnosis should be made by cardiac catheterization and angiocardiographic data. The definitive surgical management of this cyanotic malformation consists of redirection of venous blood flow toward the respective arteries by performing an intraatrial venous switch using Senning or Mustard [6] procedures.

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