

TRANSANNULAR PATCH REPAIR OF DOUBLE-OUTLET RIGHT VENTRICLE, {S,D,L}, AND SINGLE RIGHT CORONARY ARTERY

Yukihiko Kaneko, MD, Arata Murakami, MD, Kazuhito Imanaka, MD, Hideo Okabe, MD, and Shinichi Takamoto, MD,
Tokyo and Yokohama, Japan

A heart murmur and cyanosis were noted in a newborn female baby. When she was 5 years of age, cardiac catheterization and angiography revealed double-outlet right ventricle (DORV) with bilateral conus, L-malposed great arteries (that is, {S,D,L} in van Praagh's classification), single right coronary artery (RCA), and pulmonary stenosis caused by subpulmonary hypertrophied muscle and a hypoplastic pulmonary anulus. Inasmuch as the patient was free of symptoms and surgical repair was considered to entail considerable risk, repair was postponed.

At 21 years of age, because of her intention to become pregnant, the patient was referred to us again after a second cardiac catheterization and angiography. The study revealed the aforementioned anomaly and left juxtaposition of the atrial appendages. Pulmonary valve diameter was 16 mm (Z-value -2.7). The systolic pressure gradient across the pulmonary outflow tract was 83 mm Hg. An angiogram showed sufficient space at the right side of the subpulmonary conus between the pulmonary valve and the RCA (Fig 1). Cardiac repair with transannular patch enlargement of the pulmonary outflow was planned.

At surgery, a longitudinal incision was made in the right side of the main pulmonary artery and extended 4 cm into the right ventricle, equidistantly from the RCA and right atrioventricular groove (Fig 2). The minimum distance from the incision to the RCA and right atrioventricular groove was 6 mm, allowing later suturing of a patch over the incision. Subpulmonary obstructive muscle was resected, and the ventricular septal defect was connected to the aorta with a polytetrafluoroethylene gusset. The right ventricular outflow tract was augmented with a monocusp mounted polytetrafluoroethylene patch. Postoperative catheterization revealed a small residual shunt. The pulmonary/systemic flow ratio was 1.3, the right ventricular/left ventricular pressure ratio was 0.28, and the pressure gradient across the pulmonary outflow tract was 16 mm Hg. Echocardiography showed a competent

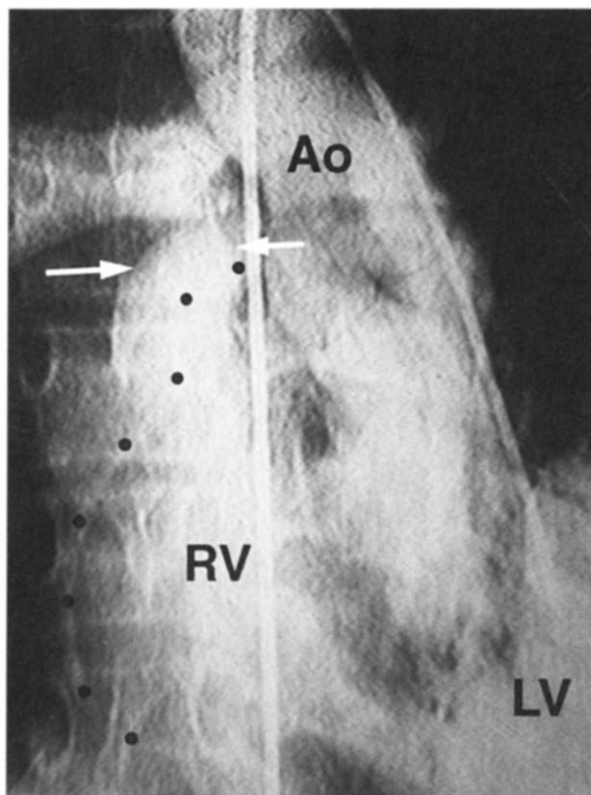


Fig 1. Preoperative left ventriculogram showed L-malposed great arteries and bilateral conus. The pulmonary anulus (white arrows) and RCA (black dots) were relatively distant at the right side of the pulmonary outflow. Ao, Aorta; LV, left ventricle; RV, right ventricle.

tricuspid valve. The patient's level of activity was better than that before the operation and she hopes to become pregnant.

L-Malposition of the great arteries associated with DORV is an infrequent combination of heart anomalies and usually coexists with pulmonary outflow tract obstruction. Otero Coto and associates¹ reported that pulmonary outflow tract obstruction was present in 73% of hearts with this combination. The anomalous position of the pulmonary valve poses difficulties with surgical relief of the pulmonary outflow tract obstruction in that the RCA crosses the pulmonary outflow tract and may hinder transannular patch placement. To our knowledge, proper positioning of a transannular patch has not been discussed in the literature.

From the Department of Cardiothoracic Surgery, University of Tokyo, Tokyo, Japan, and the Department of Thoracic and Cardiovascular Surgery, Kanagawa Children's Medical Center, Yokohama, Japan.

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Address for reprints: Yukihiko Kaneko, MD, Department of Cardiothoracic Surgery, General Hospital, University of Wien, Währinger Gürtel 18-20, 1090 Wien, Austria.

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Kirklin and Barratt-Boyes² stated that in such patients an extracardiac conduit between the right ventricle and the pulmonary artery was nearly always necessary at the time of correction. However, the location of the coronary artery in the present patient required the area of proximal conduit anastomosis to be immediately behind the sternum. Placement of either a conduit to the main pulmonary artery to the right of the aorta or a conduit to the left pulmonary artery to the left of the aorta appeared prone to conduit compression between the sternum and right ventricle.³

The REV procedure (*réparation à l'étage ventriculaire*) is another option.⁴ However, the translocated pulmonary artery might be compressed between the sternum and anteriorly displaced aorta.

Lincoln⁵ dissected out the RCA and sutured a transannular patch underneath it to enlarge the pulmonary valve ring in this anomaly. This technique stretches the RCA and thereby predisposes to coronary insufficiency. Coronary artery bypass to the distal RCA after transection of the RCA also allows transannular enlargement.³ Nevertheless, although coronary artery bypass is effective for augmenting already impaired coronary flow, it may not perfuse hypertrophied right ventricular muscle sufficiently after obliteration of the normal coronary artery.

In the present case, it was possible to relieve pulmonary stenosis properly by placing a transannular patch on the right side of the subpulmonary conus without jeopardizing the RCA. We relieved pulmonary outflow tract obstruction sufficiently by the same technique in another case with DORV, L-malposition, and a pulmonary valve Z-value of -3.0. Because the RCA arising from the levoposed aorta typically runs obliquely toward the right atrioventricular groove, there is a better chance of making a longer transannular incision at the right side of the pulmonary outflow tract than in the ventral surface. As suggested by the surgical experience with tetralogy of Fallot repair, a limited incision into the right ventricle greatly facilitates enlargement of the pulmonary anulus. Since DORV with L-malposition usually has bilateral conus and the coronary arteries are rarely located within the subpulmonary conus, transannular patch enlargement of the pulmonary anulus on the right side of the pulmonary outflow tract is probably feasible in the majority of patients with DORV and L-malposition, in whom the RCA is not adjoining the pulmonary anulus.

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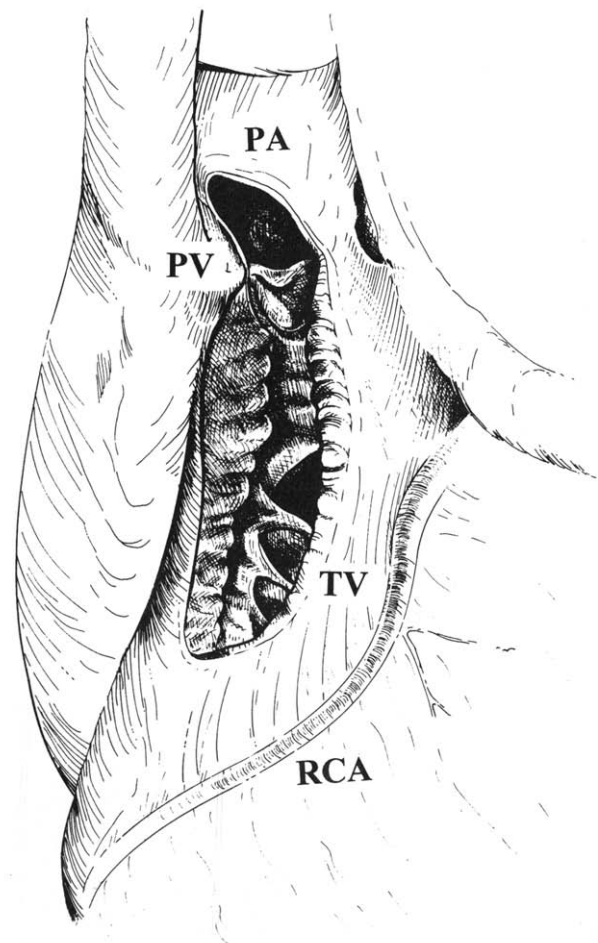


Fig 2. Drawing of the surgical view after placement of a transannular incision. The pulmonary and tricuspid valves are visualized through the incision. PA, Pulmonary artery; PV, pulmonary valve; TV, tricuspid valve; RCA, right coronary artery.

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