An arterial switch operation for a concordant crisscross heart with the complete transposition of the great arteries

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Crisscross heart is a rare cardiac malformation that results from the abnormal rotation of the ventricles.¹ We performed a successful arterial switch operation in a patient with a concordant crisscross heart that was complicated by a complete transposition of the great arteries. To our knowledge, an arterial switch operation to treat a concordant crisscross heart has not been reported.

Clinical Summary

A 2-year-old boy with a concordant crisscross heart, a complicated complete transposition of the great arteries, a ventricular septal defect (VSD), an atrial septal defect, and a patent ductus arteriosus was referred after a balloon atrioseptostomy at 1 day of age and pulmonary artery (PA) banding at 31 days of age. A chest radiograph revealed levocardia with a cardiothoracic ratio of 59%. An electrocardiogram exhibited a sinus rhythm and a mean frontal QRS axis of -70° . Preoperative cardiac catheterization showed a mean PA pressure of 18 mm Hg, a pulmonary/systemic flow ratio of 1.3, and a pulmonary vascular resistance of 2.0 Wood units (Table 1). The systolic pressures in the ventricles were identical, and the pressure gradient at the PA banding site was 63 mm Hg. Left and right ventriculograms are shown in Figure 1. An inferior ventricular injection showed an anatomically left ventricle and an intraventricular septum. The PA originated from the left ventricle. A superior ventricular injection showed an anatomically right ventricle and an ascending aorta that originated from the right ventricle. The PA was well developed, with a PA index of 406. The left ventricular end-diastolic volume was 133% of normal, with a left ventricular ejection fraction of 61%. The right ventricular end-diastolic volume was 102% of normal, with a right ventricular ejection fraction of 51%.

With the patient under fentanyl anesthesia, a median sternotomy was made. The gross anatomy was compatible with a concordant crisscross heart in Van Praagh's {S,D,L}, the right ventricular apex was toward the left, and a left anterior ascending aorta was present. Pump perfusion was established with ascending aortic and direct bicaval cannulation. During cold fibrillation, the main PA was divided at the banded site, and the right atrial appendage was opened along its summit in preparation for a modified Fontan procedure. The VSD, 15 mm in size, was located in the membranous portion of the septum, and the PA was found to originate from the left ventricle with a mitral valve-PA fibrous continuity. On the basis of these findings, an arterial switch operation seemed feasible. The VSD was closed with a Dacron polyester fabric patch with eight pledget-supported mattress sutures. After the

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Received for publication Sept 18, 2001; accepted for publication Dec 10, 2001.

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J Thorac Cardiovasc Surg 2002;124:176-8

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0022-5223/2002 \$35.00+0 12/54/122301

doi:10.1067/mtc.2002.122301

Figure 1. A, Preoperative right ventriculogram. B, Preoperative left ventriculogram. *Arrowheads* point to the mitral valve. C, Preoperative biventriculogram at VSD. *Arrowheads* point to the interventricular septum. D, Schema of preoperative anatomy. *Black broad arrow*, arterial flow; *white broad arrow*, venous flow. E, Postoperative right ventriculogram. F, Postoperative left ventriculogram. TV, Tricuspid valve; Ao, aorta; RV, right ventricle; PAB, banded site of PA; LV, left ventricle; IS, infundibular septum; IVS, intraventricular septum; LA, left atrium; RA, right atrium; MV, mitral valve.

induction of cardioplegia, the left-sided aorta was transected 15 mm distal to its origin. Both coronary arteries were translocated into the facing wall of the PA with polydioxanone suture. After the French maneuver, an end-to-end anastomosis was made between the distal aorta and the main PA stump. The coronary defect in the aortic root was closed with a pericardial patch with polydioxanone suture. PA continuity was reestablished by an anastomosis between the PA bifurcation and the old aorta stump with polydioxanone suture. The patient did well after the operation and was discharged from the hospital on the 21st postoperative day. A postoperative cardiac catheterization was performed 6 months after the operation. The findings are shown in Table 1. Postoperative left and right ventriculograms are shown in Figure 1. The left and right ventricular volumes remained the same as before the operation.

Discussion

Only two cases of definitive biventricular repairs^{2,3} and a few cases of modified Fontan procedures^{4,5} for the surgical treatment of

concordant crisscross heart have been reported. We have performed two biventricular repairs,² including this case, and 17 modified Fontan procedures in patients with concordant crisscross heart anomalies from June 1983 to March 2001. The 17 modified Fontan procedures were indicated for hypoplastic right ventricles ($n = 3$), hypoplastic left ventricles ($n = 2$), straddling atrioventricular valves ($n = 4$), and ventricular anatomic features ($n = 8$). In the 8 patients with ventricular anatomic features, VSDs were present that occupied almost the entire inflow of the right ventricle and reached its apex, where the chordae of the mitral valve were observed. Among the 17 modified Fontan procedures, 1 late death occurred (of congestive heart failure at 13.8 months after the operation).

In the case of a concordant crisscross heart with a complicated straddling of the atrioventricular valve, a biventricular repair is not suitable. Such cases require a modified Fontan procedure. Important considerations in deciding to adapt a biventricular repair are the presence of hypoplastic one ventricle, a straddling of the atrioventricular valve, and the other ventricular anatomic features.

TABLE 1. Preoperative and postoperative cardiac catheterization data, ventricular volumes, and ejection fraction

Site	Before operation		After operation	
	Pressure (mm Hg), max/min (mean)	Oxygen sat (%)	Pressure (mm Hg), max/min (mean)	Oxygen sat (%)
SVC	(4)	63		
IVC	(4)	61		
RA	(4)	70	(4)	78
RV	85/-, EDP 7	71	28/-, EDP 4	77
MPA	22/12 (18)	87	24/8 (16)	77
PAW			(8)	96
PV	(9)	97		
LA	(7)	97		
LV	85/-, EDP 7	92	90/-, EDP 8	95
Ao	85/55 (68)	93	90/53 (72)	95
LVEDV	133% of normal		129% of normal	
LVEF	0.61		0.56	
RVEDV	102% of normal		134% of normal	
RVEF	0.51		0.53	
Qp	5.4			
Qs	4.1			
Qp/Qs	1.3			
Rp	2.0			
Rs	15.6			

SVC, Superior vena cava; IVC, inferior vena cava; RA, right atrium; RV, right ventricle; MPA, main pulmonary artery; PAW, pulmonary artery wedge pressure; PV, pulmonary vein; LA, left atrium; LV, left ventricle; Ao, aorta; sat, saturation; Max, maximum; Min, minimum; LVEDV, left ventricular end-diastolic volume; LVEF, left ventricular ejection fraction; RVEDV, right ventricle end-diastolic volume; RVEF, right ventricular ejection fraction.

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