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Brief Report

Double switch operation in a young infant

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Abstract We performed a combined Senning and arterial switch operation on a 2-month-old patient with congenitally corrected transposition, Ebstein's malformation producing severe tricuspid regurgitation, ventricular septal defect, pulmonary hypertension, and congestive heart failure. The tricuspid regurgitation was improved. The double switch operation has the advantage of improving the function of the systemic atrioventricular valve, especially in newborns or young infants in whom the outcome of the valvar repair is poor.

Keywords: Anatomical repair; arterial switch operation; Senning procedure; congenitally corrected transposition; Ebstein's malformation

THE DOUBLE SWITCH OPERATION, COMBINING THE Senning and arterial switch procedures, is reported to have excellent results over the short and mid term as an anatomical repair for patients with congenitally corrected transposition, and it has been suggested that it can prevent future failure of the systemic ventricle and atrioventricular valve.[1–3] Although several discussions have focused on the risks of this procedure in older children, little attention has been given to its indication in the newborn and young infant. As far as we know, very few accounts have been given for use of this procedure in the newborn or young infant less than 3 months old.[2–4] We have recently performed the procedure in a young infant with congenitally corrected transposition, Ebstein's malformation, ventricular septal defect, and pulmonary hypertension. In this report, we describe and discuss our experience.

Case report

A 2-month-old boy weighing 4600 grams was noted to have a heart murmur at the age of 1 month. At our hospital, we made the echocardiographic diagnosis of congenitally corrected transposition, ventricular septal defect, small atrial septal defect, and Ebstein's malformation producing severe tricuspid regurgitation (Fig. 1a). We followed the patient as an outpatient, treating him with an inhibitor of angiotensin converting enzyme, but he was soon re-admitted with congestive heart failure. Despite medical treatment, the tricuspid regurgitation progressively became worse, and the patient required mechanical ventilation. Cardiac catheterization confirmed the echocardiographic diagnosis, confirming the presence of pulmonary hypertension, and left and right ventricular volume overload (Table 1).

We proceeded, therefore, to surgery on a semi-emergency basis using cardiopulmonary bypass with moderate hypothermia. Under ventricular fibrillation, the right atrium was entered through an incision made parallel to, but 12 millimetres away, from the terminal groove. The atrial septal defect was closed directly, and the septal flap was developed. The tricuspid valve showed evidence of Ebstein's malformation, with downward displacement of the attachments of the septal and mural leaflets. The atrialized ventricle was plicated longitudinally along the mural leaflet with 5-0 polypropylene mattress sutures. The Senning operation was performed mostly under ventricular fibrillation. The ventricular septal defect was of a muscular outlet type in the subpulmonary position. The pulmonary artery trunk was transected at the bifurcation, and the ventricular septal defect was closed in transpulmonary fashion. The anterior atrioventricular conduction was preserved as suggested by de Leval et al.[5] Then an arterial switch operation was performed using the Lecompte modification and the trapdoor technique. The old aortic root was enlarged

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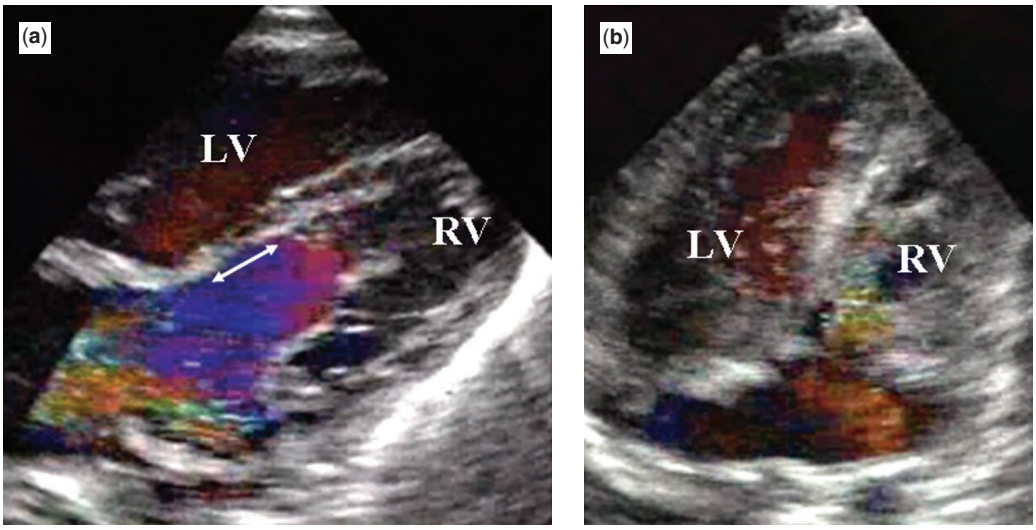

Figure 1.
*Transthoracic echocardiogram. (**a**) Pre-operative view showing severe tricuspid regurgitation and atrialization of the morphologically right ventricle. (**b**) The post-operative scan shows mild tricuspid regurgitation. LV: morphologically left ventricle; RV: morphologically right ventricle; dual arrow: atrialized right ventricle.*

Table 1. Cardiac parameters.

	Pre-operation	Post-operation
LVEDV (% of normal)	224	169
LVEF (%)	66	52
RVEDV (% of normal)	201	117
RVEF (%)	66	54
Systolic LVp (mmHg)	58	82
Systolic RVp (mmHg)	55	25
TR	Severe	Mild

Abbreviations: mmHg: millimetres of mercury; %: per cent; LVEDV: morphologically left ventricular end-diastolic pressure; LVEF: morphologically left ventricular ejection fraction; RVEDV: morphologically right ventricular end-diastolic pressure; RVEF: morphologically right ventricular ejection fraction; LVp: morphologically left ventricular pressure; RVp: morphologically right ventricular pressure; TR: morphologically tricuspid valvar regurgitation

with autologous pericardium. Cardiopulmonary bypass lasted for 297 minutes, with 155 minutes of aortic occlusion.

The post-operative clinical course was uneventful. Cardiac catheterization at 8 months after the operation showed marked reduction in both left and right ventricular volumes, and normal pressures in the morphologically right ventricle (Table 1). Tricuspid regurgitation, as seen on the echocardiogram, was decreased from severe to mild (Fig. 1b).

Discussion

The double switch operation, as performed in our patient, successfully improved congestive heart failure due to tricuspid regurgitation and excessive pulmonary blood flow. The procedure, nonetheless, has rarely been performed in small children, so careful consideration is needed prior to embarking on this operative course. Karl et al.[4] suggested that neonates with symptoms due to excessive flow of blood to the lungs in the presence of a large ventricular septal defect should undergo banding of the pulmonary trunk in preparation for a definitive operation at about 12 months of age, but that the double switch operation should be performed irrespective of time if the condition of the patient becomes complicated by severe tricuspid regurgitation and morphologically right ventricular dysfunction.[4] Nine-tenths of cases with congenitally corrected transposition are known to have structural abnormalities of the morphologically tricuspid valve, such as dysplasia of the leaflets, and thickened cordal attachment of the septal and mural leaflets,[6] and at least one-tenth of them have an Ebstein-like valve.[7] Repair is difficult in this context, the outcome being poor because of the limited surgical vision due to the location of the morphologically right ventricle.[7,8]

In patients with congenitally corrected transposition, tricuspid regurgitation and congestive heart failure, but normal morphologically left ventricular pressure, banding of the pulmonary trunk may improve tricuspid regurgitation because of the shift of the ventricular septum due to the increase in left ventricular pressure.[9] For these patients, banding in preparation for the double switch operation, as opposed to tricuspid valvar repair, is a good surgical option because this strategy permits observation of the progress of the tricuspid regurgitation, and retains the option of the double switch operation. In the patient

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who already has systemic pressure in the morphologically left ventricle, however, the effect of banding on tricuspid regurgitation is uncertain. In our patient, tricuspid regurgitation was improved post-operatively by simple tricuspid annuloplasty and plication of the atrialized morphologically right ventricle, with the tricuspid valve then converted to a low-pressure environment. This is a significant advantage of the double switch operation, especially in the newborn or young infant in whom repair of the tricuspid valve is difficult. The late sequels of this procedure as performed in newborns and young infants are unknown, but we now know that the procedure is feasible. Thus, we suggest that the double switch operation can be a procedure of choice in the newborn or young infants presenting with congestive heart failure due to systemic atrioventricular valvar regurgitation in the setting of congenitally corrected transposition.

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