Symptomatic Aortic Regurgitation after Blalock-Taussig Shunt in Tetralogy of Fallot with Bicuspid Aortic Valve

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—— We report here a case of a premature baby with tetralogy of Fallot and
bicuspid aortic valve. After the successful completion of the Blalock-Taussig
(BT) shunt, severe aortic valve regurgitation (AR) appeared, although it was
trivial preoperatively. Severe postoperative heart failure was induced by progres-
sion of the AR. Postoperative echocardiography revealed that the progression of
the AR was provoked by appearance of prolapse of the cusp as the result of rapid
increase of blood flow through the aortic valve after the BT shunt. We propose
that, in planning the BT shunt for patients with tetralogy of Fallot, preoperative
examinations for a possible bicuspid aortic valve should be done and postoperative
precaution considering possible appearances of severe AR and congestive heart
failure will be necessary. ——— tetralogy of Fallot; bicuspid aortic valve;
aortic regurgitation; Blalock-Taussig shunt © 1997 Tohoku University Medical
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The association of tetralogy of Fallot with malformations of the aortic valve is uncommon. Most of the association of the aortic valve in adults and older children is aortic valve regurgitation (AR) induced by deformity of the cusps as a result of bacterial endocarditis and by severe dilatation of the aortic annulus and right coronary cusp prolapse as a late complication (Bahnson et al. 1962; Lev and Eckner 1964; Glancy et al. 1968; Beach et al. 1971; Peters 1971; Rao et al. 1971; Rieker et al. 1975; Matsuda et al. 1980). We present here rare case of tetralogy of Fallot associated with bicuspid aortic valve who developed severe AR and severe congestive heart failure after Blalock-Taussig (BT) shunt. We stress the clinical importance of this association and discuss the possible mechanism of AR in bicuspid aortic valve.

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CASE REPORT

A female infant was born at 26 weeks of gestation weighing 909 g. She showed a mild cyanosis since birth and was diagnosed as having extreme tetralogy of Fallot with patent ductus arteriosus and trivial AR by two-dimensional and color Doppler echocardiographies. Her ventricular septal defect was located in the subaortic area and did not extend to the subpulmonic area. After the age of 67 days cyanosis gradually became severe due to the constriction of the ductus arteriosus, and a continuous intravenous infusion of prostaglandin E1 (PGE1) was started. Cardiac catheterization and angiography were performed at 79 days of age. There was no systolic pressure gradient across the aortic valve. The angiography revealed pulmonary atresia and patent ductus arteriosus (Fig. 1). The diameters of both right and left pulmonary arteries were 3 mm, and those of subclavian arteries were both 2 mm. As the response to PGE1 gradually became poor, the right original BT shunt procedure was palliatively performed at 111 days of age with 3.5 kg of body weight. Ligation of the ductus arteriosus was not performed. Although her cyanosis was improved after the shunting and the

Fig. 1. Preoperative finding of aortography. Pulmonary atresia and patent ductus arteriosus were detected. AO, aorta; DA, ductus arteriosus; R(L)PA, right (left) pulmonary artery.

arterial oxygen tension arised from 31 to 42 torr, severe congestive heart failure gradually progressed from the 5th postoperative day, and a marked right lung congestion and a cardiomegaly were found on chest roentgenograms. The cardiothoracic ratio increased from 55 % to 68 %. Diastolic murmurs were heard near the aortic valve. Severe degree of AR reaching to the tip of mitral valvular leaflet was detected by color-Doppler and contiuous Doppler echocardiographies (Fig. 2). The absence of the right coronary-noncoronary commissures, i.e., a bicuspid aortic valve with a combined right coronary-noncoronary leaflet was

Fig. 2. Postoperative findings of Doppler echocardiography. Severe degree of aortic valve regurgitation was detected.

Fig. 3. Two-dimensional echocardiographic finding of aortic valve in short axis view. A bicuspid aortic valve with a combined right coronary-noncoronary leaflet was revealed (arrow). AV, aortic valve; LA, left atrium.

revealed by two-dimensional echocardiography (Fig. 3). Comparing with the preoperative findings, postoperative left ventricular end-diastolic dimension, left atrial dimension, and velocity time integral of aortic flow detected by pulsed-Doppler method were all increased (Table 1), and the prolapse of the right coronary-noncoronary cusp was newly detected (Fig. 4). By the 10th day after the operation, both congestive heart failure and hemodynamics were alleviated and stabilized by a drastic restriction of water intake and by administrations of catecholamines and diuretics in addition to the closure of the ductus arteriosus occurred at the 7th day after the operation. However, the prolapse of the cusp and severe AR remained inspite of the closure of ductus arteriosus. Because of the cardiomegaly on chest roentgenogram and mild symptoms of heart failure due to the remained severe AR, administration of diuretics has been unable to be discontinued for 6 months after the operation.

TABLE 1. *Preoperative and postoperative findings of echocardiography*

	Pre BT shunt	Post BT shunt
Heart rate (beat/min)	125	147
Left ventricular end-diastolic dimension (cm)	1.3	1.6
Left ventricular shortening fraction	0.31	0.30
Left atrial dimension (cm)	1.1	1.5
Aortic valve annular dimension (cm)	0.9	1.0
Prolapse of right coronary-noncoronary cusp	(-)	(+)
Aortic regurgitation	Trivial	Severe
Time velocity integral of aortic flow (cm)	14.9	29.6

Fig. 4. Postoperative finding of echocardiography. The prolapse of the right coronary-noncoronary cusp are seen (arrow). AO, aorta; RV, right ventricle; LV, left ventricle.

DISCUSSION

AR caused by bicuspid aortic valve associated with tetralogy of Fallot is uncommon (2%) (Glancy et al. 1968). To our knowledge, our patient is the first infantile case of AR progressed rapidly after BT shunt. Van Praagh and McNamara (1968) described 2 cases of ventricular septal defect of the tetralogy type associated with bicuspid aortic valve and AR. They concluded that AR in their cases was induced by downward prolapse of the right coronary-noncoronary cusp because of the lack of support for the cusp from below by the conal musculature and abnormal cusp apposition in diastole resulting from bicuspid aortic valve. We think the mechanism of AR in our case as follows. Our patient had bicuspid aortic valve and congenital abnormality of the coaptation of the bicuspid leaflets. In addition to such morphological abnormalities of aortic valve, the rapid increase of blood flow through the aortic annulus after the BT shunt produced volume overload on the aortic valve. As a result, the downward prolapse of right coronary-noncoronary cusp appeared, which made the apposition of the bicuspid leaflets poor, leading to severe and irreversible AR. Although the mechanism of AR in isolated bicuspid aortic valve is unclear (Roberts 1970; Stewart et al. 1984), our case indicates that prolapse of cusp relates with the mechanism of AR in tetralogy of Fallot associated with bicuspid aortic valve. Our experience with the present patient suggests that before BT shunt for tetralogy of Fallot, examinations for aortic valve abnormalities are essential and echocardiography seems to be the best procedure for this purpose. When a bicuspid aortic valve is detected, postoperative management should be considered against the development of AR and congestive heart failure. As in our case, water intake will be critical; its sufficient volume is needed to maintain the perfusion of BT shunt, while its excessive volume can cause AR and heart failure. In addition, a simultaneous ligation of the ductus arteriosus will be effective to prevent excessive increase of blood flow through the aortic valve. We suppose that surgical treatment for the aortic valve of this patient should be considered when the intracardiac repair will be planned in future.

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