DOUBLE OUTLET RIGHT VENTRICLE WITH SEVERE LEFT VENTRICULAR
OUTFLOW TRACT OBSTRUCTION DUE TO SMALL VENTRICULAR
SEPTAL DEFECT AND ANOMALOUS ADHERENCE OF THE
MITRAL VALVE TO THE VENTRICULAR SEPTUM

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A 1-month-old male infant with a double outlet right ventricle complicated
by left ventricular outflow tract obstruction due to a very small ventricular
septal defect is described. The atrial septum was intact, but mitral stenosis
was present. The characteristic finding in this patient was that the inherently
small ventricular septal defect was further narrowed by abnormal adherence
of the anterior mitral leaflet to the left ventricular septal surface below the
defect. Intracardiac anatomical features other than the adherent mitral
valve could be evaluated accurately by two-dimensional echocardiography.
Because the atrial septum was intact, balloon atrioseptostomy was not possi-
ble. An attempt at surgical creation of an atrial septal defect was unsuccess-
ful, and the patient died immediately after the operation. The autopsy
findings of the heart are described, and diagnostic problems and the possibility
of radical operation are discussed.

The double outlet right ventricle is an un-
common congenital malformation in which
both the pulmonary artery and the aorta origi-
nate entirely from the right ventricle. A ven-
tricular septal defect is necessary for survival,
since it represents the only outlet of the left
ventricle. The size of the ventricular septal defect
is critical; too small a defect leads to obstruction
of the left ventricular flow and left heart
failure!—13 Since a double outlet right ventricle
with obstruction of the left ventricular outflow
tract due to small ventricular septal defect may
be treated successfully by enlargement of the
ventricular septal defect, early detection of the
condition is important. There have been few
reports of a double outlet right ventricle ac-
accompanied by left ventricular outflow tract
obstruction being diagnosed during life2—7,11—13
but in all these cases the diagnosis was made by
cardiac catheterization and angiocardiography.
In a 1-month-old male herein described, a double
outlet right ventricle accompanied by mitral
stenosis and a small ventricular septal defect was
diagnosed by two-dimensional echocardiography.
The patient died immediately after surgical
creation of an atrial septal defect. In addition to
echocardiographic findings, autopsy revealed a
rare condition; the small ventricular septal defect
was further narrowed by abnormal adherence of
the anterior mitral leaflet to the left ventricular
septal surface below the defect.

CASE REPORT
The patient was born after full term gestation

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with a weight of 3,530g. His mother noted cyanosis of the lips during crying about 1 week after birth. Intake of the breast milk was slightly reduced. Peripheral cyanosis and heart murmur were noted at a check-up 1 month after birth. The boy was admitted to the Department of Pediatrics, Miyazaki Medical College Hospital, for further evaluation. On admission, the infant was 54 cm tall and weighed 4.1 kg with a regular pulse rate of 100/min. Tachypnea of 50 breaths per minute with subcostal retraction but no rales were heard over the chest. Mild central cyanosis was observed. A pansystolic murmur, grade 3/6, was heard along the lower left sternal border.

The second heart sounds were shortly split, and pulmonary sound was increased. The liver was palpable 1 cm below the right subcostal margin on the midclavicular line. The chest roentgenogram (Fig. 1, top) showed an enlarged right ventricle and main pulmonary artery, and the cardiothoracic ratio was 57%. Pulmonary vascular markings were not striking and signs of pulmonary congestion were absent. An electrocardiogram (Fig. 1, bottom) revealed sinus rhythm and right axis deviation (AQRS = 150°). Positive T waves in the right precordial leads and deep S waves in the left precordial leads indicated right ventricular hypertrophy, and inverted T waves in

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DORV with Severe LVOT Obstruction

Fig. 2. Two-dimensional echocardiogram and M-mode echocardiogram. Left: an apical four-chamber view showing a small slit-like ventricular septal defect below the anterior mitral leaflet (white arrowhead) and a dilated left ventricle. Right: M-mode features of the anterior mitral leaflet showing reduced excursion and displacement toward the ventricular septum. RA = right atrium, LA = left atrium, RV = right ventricle, LV = left ventricle, IVS = interventricular septum.

Fig. 3. Right ventriculogram. A and B are simultaneous posteroanterior and left lateral projections, respectively. Ao = aorta, PA = pulmonary artery.

the left precordial leads suggesting a marked pressure overload of the left ventricle. Two-dimensional echocardiography showed bilateral coni of the aorta and pulmonary artery, both originating from the right ventricle. The two great arteries were positioned nearly side by side, but the aorta was to the right and slightly posterior to the pulmonary artery. Although not distinctly seen in stop-frame pictures, an apical four-chamber view showed a small ventricular septal defect below the anterior mitral leaflet (white arrowhead), a spherically dilated left ventricle with reduced contractility, and an enhancement of echo signals on the left ventricular endocardial surface. The atrial septum was intact (Fig. 2, left). The mitral valve was anteriorly displaced toward the ventricular septum and both anterior and posterior mitral leaflets showed small excursion with a reduced motion range, suggesting mitral stenosis (Fig. 2, right). M-mode echocardiography of the pulmonary valve suggested pulmonary hypertension. The patient was diagnosed by two-dimensional echocardiography as having double outlet right ventricle accompanied by obstruction of the left ventricular outflow tract and pulmonary hyper-
Fig. 4. Heart specimen. A: Opened right ventricle (RV) to show the aorta (Ao) originating above hypertrophied RV. A small slit-like ventricular septal defect (VSD) is observed below the aortic valve (black arrow). B: Left atrium and left ventricle (LV) opened to show the course of probe from RV through VSD; endocardial fibroelastosis; inherently small VSD nearly obstructed by the anterior mitral leaflet connected with a chorda adhering to the left ventricular septal surface below the VSD; chorda of posterior mitral leaflet showing anomalous adherence to the left ventricular lateral free wall; thickened and stenotic mitral valve. C: RV opened to show "the right ventricle-pulmonary compartment"; absent pulmonary stenosis. D: Opened right atrium (RA) to show intact atrial septum; enlarged and hypertrophied RA.

tension due to a small ventricular septal defect complicated by mitral stenosis. After admission, cyanosis increased, the feeding of milk was decreased and sporadic attacks of bradycardia began to occur. Since the condition was considered to be critical with marked obstruction of the left ventricular outflow tract, cardiac catheterization and angiocardiography were performed for definitive diagnosis and balloon atrioseptostomy. The catheter was advanced directly

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from the right ventricle into the aorta and pulmonary artery. The systolic pressure was identical in the right ventricle and the two great arteries. Step up of oxygen saturation was 8% in the right ventricle and 13% in the aorta. Blood gas analysis showed pH 7.35, PaO₂ 23.5 mmHg and PaCO₂ 41.4 mmHg. There was no interatrial communication, and the catheter could not be inserted into the left atrium or the left ventricle. Balloon atrioseptostomy was impossible for the same reason. In right ventriculograms (Figs. 3A and 3B), both the aorta and pulmonary artery had bilateral conl originating from the right ventricle. Opacification of the left ventricle due to right-to-left shunt through the ventricular septal defect was not possible. In the levophase of right ventriculograms, a spherically dilated left ventricle with markedly reduced contractility was observed. Although the left-to-right shunt flow through the ventricular septal defect could not be identified, slight reopacification was noted in the aorta. The disappearance of the contrast material from the left ventricle was markedly delayed. An attempt at surgical creation of an atrial septal defect with a small sharp blade on the 50th day of life employing no pump-oxygenator was unsuccessful, and the patient died immediately after the operation despite intensive efforts to resuscitate. Autopsy revealed that both the aorta and pulmonary artery originated from the right ventricle, and were of identical size. The two arteries were positioned nearly side by side, but the aorta was located to the right and slightly posteriorly to the pulmonary artery. Marked hypertrophy and dilatation of the right atrium, right ventricle and left ventricle were seen. The left atrium was hypertrophied but not dilated. A ventricular septal defect was observed below the aortic valve, and measured only 3 x 2 mm (Fig. 4A). The mitral valve orifice was small and the anterior mitral leaflet was thickened, resulting in restriction of the motion of the valve as in mitral stenosis. In addition, the small ventricular septal defect was nearly obstructed by the anterior mitral leaflet, connected with a shortened chordae anomalously attached to the left ventricular septal surface below the defect. The left ventricular endocardium was thickened, showing marked endocar- dial fibroelastosis. The finding of endocardial fibroelastosis indicates a strong pressure overload exerted on the left ventricle during life due to stenosis of the left ventricular outflow tract (Fig. 4B). Pulmonary or subpulmonary stenosis was not observed (Fig. 4C). The foramen ovale was not patent, and the atrial septal defect was not well made (Fig. 4D).

DISCUSSION

Double outlet right ventricle with obstruction of the left ventricular outflow tract due to a small ventricular septal defect was first reported by Edwards et al! in 1952, and, to our knowledge, 20 similar cases have been documented to date!-13 Among studies employing a relatively large number of patients, double outlet right ventricle with a stenotic ventricular septal defect was reported in 3 (5.0%) of 62 patients by Stridarmont et al!! 4 (17.3%) of 23 patients by Zamora et al!0 and 3 (17.6%) of 17 patients who underwent operation by Kirklin et al!.

In double outlet right ventricle, malalignment of the truncoconal septum and primitive ventricular septum usually results in a large ventricular septal defect!° When the two are well aligned, the ventricular septum becomes intact, but when there is incomplete alignment, a small ventricular septal defect may persist. In some patients, the inherently small ventricular septal defect may be further narrowed by hypertrophy around the defect due to a jet lesion13 or by surrounding endocardial cushion tissue!° In our patient, an originally small ventricular septal defect was obstructed by abnormal adh- erence of the anterior mitral leaflet to the left ventricular septal surface below the defect. An analogue to this unique condition has been reported in only 1 patient by Edwards et al.

The development of left heart failure and anginal pain as well as marked left ventricular hypertrophy rarely seen in electrocardiograms of ordinary cases of double outlet right ventri- cle!° are among the clues to a clinical diagnosis of obstruction of the left ventricular outflow tract associated with double outlet right ventricle due to a small ventricular septal defect. This condition has been diagnosed definitively by examining the systolic pressure gradient between the left and right ventricles by cardiac catheterization and angiographic evaluation of the ventricular septal defect. These invasive methods were employed in all previous cases in whom the condition was diagnosed during life2-7,11-13 However, cardiac catheterization in double outlet right ventricle accompanied by a small ventricular septal defect involves

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considerable risk, and sudden death during or immediately after examination has been reported in 5 patients\textsuperscript{3,5,7,9,13} In our patient, the intracardiac anatomy other than the anomalous adherence of the chorda supporting the anterior mitral leaflet to the left ventricular septal surface could be evaluated accurately by two-dimensional echocardiography. The adherent mitral valve may be directly visible in some cases if the resolution of the apparatus is sufficient. The anterior mitral leaflet displaced toward the ventricular septum and showing a reduced excursion and motion range observed in two-dimensional echocardiography suggested mitral stenosis due to the adherent mitral valve.

In patients with a double outlet right ventricle with severe stenosis of the left ventricular outflow tract who develop congestive heart failure in the neonatal period to early infancy,\textsuperscript{7,8} balloon atrioseptostomy based on early diagnosis is the only treatment to prolong life. Radical surgical treatment may become possible if the left ventricular outflow tract is not severely obstructed and the patient has grown out of infancy. The operation involves incision for enlargement of the upper margin of the ventricular septal defect and construction of an intracardiac tunnel by connecting the defect with the aortic valve orifice using a patch. This procedure has been successful in 5 patients\textsuperscript{4,6,7,12,13} Since our patient presented with additional difficult problems such as left ventricular endocardial fibroelastosis and obstruction of the defect by the adherent mitral valve, the possibility of radical operation was considered to have been small even if creation of the atrial septal defect had been successful.

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