Long-Term Survival in Double Inlet Left Ventricle Combined With Pulmonary Stenosis and Parachute Mitral Valve

A Rare Case

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SUMMARY

Univentricular heart is a rare and complex congenital disease. Its prognosis is usually poor without surgical intervention and long-term survival into adulthood is rare. We report a 41-year-old man with a double inlet left ventricle with pulmonary stenosis and a parachute mitral valve verified by echocardiography and magnetic resonance imaging (MRI). (Int Heart J 2007; 48: 261-267)

Key words: Single ventricle, Univentricular heart, Pulmonary stenosis, Mitral valve stenosis, Parachute mitral valve

UNIVENTRICULAR heart or single ventricle is a congenital defect that is defined as the presence of a single ventricular chamber or 2 ventricular chambers in which one of the ventricles is severely hypoplastic. Other problems associated with single ventricle are pulmonary stenosis or atresia, subaortic stenosis, coarctation of the aorta, and mitral or tricuspid atresia.1) Therefore, many anatomical subtypes and variations exist. Neonates with functional single ventricles have a very high morbidity and mortality, making them candidates for surgical correction. Patients who have not undergone corrective surgery have a median survival of 14 years.2) Survival into late adulthood, without surgical repair, is rarely reported.3,4)

We report the case of a 41-year-old man who has a double inlet left ventricle with pulmonary atresia and a parachute mitral valve based on the echocardiographic and MRI findings.
CASE REPORT

A 41-year-old man was admitted to GyeongSang National University Hospital with the complaints of worsening dyspnea and palpitations. During childhood a cardiac disease was suspected because of mild dyspnea and exercise intolerance. He underwent cardiac catheterization and angiography at 19 years of age, and complete diagnosis was established. However, he refused any surgery and was lost to follow-up. As of the present time, he has suffered dyspnea and is classified as having NYHA class II-III functional capacity. On admission, physical examination showed moderate lip cyanosis with finger clubbing. At rest, blood pressure was 90/50 mmHg, and heart rate was about 70 beats/min and irregular. Auscultation revealed a loud diastolic murmur, heard most distinctly at the left upper sternal border and the apex. The lung sounds were clear. Mild hepatomegaly and pretibial pitting edema were found.

His laboratory findings were as follows: oxygen saturation 86.9%, hemoglobin 20.3 g/dL, hematocrit 57%, platelet count $117 \times 10^3$ mm$^3$, ALP 97 IU/L, AST 23 IU/L, ALT 13 IU/L, total bilirubin 1.4 mg/dL, uric acid 10.0 mg/dL, BUN 16.0 mg/dL, Cr 1.0 mg/dL, UA with micro: clear, SG: 1.020, protein: TR, urobilinogen: TR, bilirubin: TR, blood: -, RBC: 0 to 1, WBC: 5 to 9, epithelial cells, 5 to 9, hyaline cast: many. Electrocardiography showed atrial fibrillation, right axis deviation, and left ventricular hypertrophy. Chest radiography showed an

Figure 1. MR images of the single ventricle.
A: The 4-chamber view demonstrates a single double-inlet ventricle with intact interatrial septum as indicated by the arrow. This view clearly shows the severe mitral stenosis(*).
B: The coronal view shows the bulboventricular foramen, as indicated by the arrow, connects the left single ventricle and rudimentary right ventricle.

An aorta arose from the rudimentary right ventricle without obstruction. A right-sided pulmonary artery originated from the morphological LV and connected to the common ventricle.

SV indicates single ventricle; LA, left atrium; RA, right atrium; Ao, aorta; PT, pulmonary trunk; and rRV, rudimentary right ventricle.
enlarged heart, widened mediastinum, and pulmonary venous congestion due to mitral stenosis. Transthoracic and transesophageal echocardiography and magnetic resonance imaging revealed 2 atria with an intact interatrial septum, 2 atrioventricular valves connected to a single, morphologically intact left ventricle, and a rudimentary right ventricle which was located anteriorly. An aorta arose from the rudimentary right ventricle without obstruction. A right-sided pulmonary trunk originated from the morphological LV and connected to the common ventricle (Figure 1). The pulmonary trunk was hypoplastic and stenotic. Small, multiple collateral vessels from the aorta to the pulmonary trunk were found on MRI (Figure 2). Also, a parachute mitral valve that caused severe mitral stenosis

**Figure 2.** Relationship between great arteries and presence of pulmonary stenosis.

A: The axial view demonstrates the transposition of the great arteries (ventriculoarterial discordance). The aortic root originated from the rudimentary right ventricle, and is oriented left and slightly anterior of the pulmonary trunk (L-transposition). The pulmonary trunk originated from the SV.

B: The pulmonary trunk is hypoplastic and stenotic as indicated by the arrow.

C: The sagittal view shows pulmonary stenosis and small, multiple collateral vessels from the aorta to the pulmonary trunk, indicated by the arrow.

Ao indicates aorta; PT, pulmonary trunk; and SV, single ventricle.
was detected (Figure 3) and on transesophageal echocardiography, a left atrial appendage and a left atrial mural thrombus were seen (Figure 4).
DISCUSSION

A parachute mitral valve is a rare congenital anomaly in which shortened chordae tendinae converge and insert into a single papillary muscle. It is further characterized by thickened valve leaflets with shortening and fusion of chordae tendinae, accessory mitral valve tissue, and a supravalvular circumferential ridge of connective tissue. Ninety-nine percent of patients had associated cardiac anomalies. An isolated anomaly is rare. A double inlet left ventricle was present in 14%. The characteristic feature of an atrioventricular valve malformation is the convergent left ventricular chordal insertion into either one or 2 foci of insertion, instead of the normal divergent insertion into 2 well-separated foci.

The embryology of a double inlet left ventricle with a parachute mitral valve is incompletely understood. Van Mierop suggested that the structure separating the 2 chambers is homologous with the major part of the normal ventricular septum and that it is displaced to the right. This it was stated occurs when the AV canal is abnormally oriented, becoming “trapped” to the right AV ostium, the septum is pushed rightward and blends with the trabecula septomarginalis, while the posterior medial muscle bundle cannot blend with the posterior portion of the muscle septum. This posterior ridge that formed concomitant with partitioning of the AV canal does retain its normal relationship with the atrial septum, crux cordis, and the posterior descending artery. The mode of atrioventricular connection
seems determined by events taking place in the septation of the AV canal. If the endocardial cushions develop normally, then the canal will be divided, resulting in a double inlet. A later abnormality may result if one of these orifices becomes imperforate. If the partitioning fails to occur, a common AV valve results. If partitioning occurs asymmetrically, stenosis of one orifice will result.

Draulans-Noë reported a correlation between the type of atrioventricular valve malformation and degree of development of the lateral leaflet and accessory orifice with its tension apparatus in anatomical specimens.8)

Most patients with a single ventricle suffer from dyspnea, cyanosis, and reduced exercise tolerance. Survival is only 30% for the first year of life.10) The clinical presentation and prognosis depend on the presence or absence of an obstruction to pulmonary blood flow and the level of pulmonary vascular resistance.3) If severe pulmonary stenosis is present, it will lead to severe oxygen desaturation. Most of these patients either receive surgical treatment or die during infancy. Moderate pulmonary stenosis leads to little or no heart failure, mild cyanosis, and no pulmonary arteriolar disease.

All of the reported long-term survival cases were of the left-type morphology. The presence of discrete atrioventricular valves and adequate valvular function is associated with a better prognosis.11) In our case, long-term survival of this patient was likely achieved because of a left-type ventricle with a balanced hemodynamic condition with moderate pulmonary stenosis and moderate mitral stenosis. By the 5th decade of life, pulmonary stenosis has gradually progressed resulting in multiple collateral vessels from the aorta and its branches to pulmonary arteries, and mitral stenosis has also gradually progressed resulting in atrial fibrillation and worsened dyspnea by pulmonary venous congestion. The present findings suggest that a balanced hemodynamic condition can allow patients to live into adulthood even if they choose to forego an operation. The single ventricle requires early recognition and identification of complex defects for successful palliative surgical procedures. In the present case, the indication for late surgical repair is debatable because of a significant risk of mortality and late morbidity. This poses a dilemma, because if his condition deteriorates, he may no longer be an ideal candidate for a Fontan-type operation. Hence, close follow-up is essential, with careful clinical evaluation, periodic echocardiography, and exercise stress testing. The patient received standard heart failure treatment consisting of angiotensin converting enzyme inhibitor and beta-blocker administration. Also, he was started on low-dose diuretics to reduce pulmonary congestion. We choose digitalis for control of heart rate and an anticoagulant to prevent thromboembolism. We have discussed a double inlet left ventricle with pulmonary stenosis and a parachute mitral valve in a 41-year-old man who has not undergone surgical intervention.
REFERENCES


