Staged Fontan’s operation for unguarded tricuspid orifice with pulmonary atresia

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Abstract

We report a rare case of unguarded tricuspid orifice with pulmonary atresia, which is devoid of any guarding valvular tissue at tricuspid annulus. A left Blalock—Taussig shunt was constructed on the 17th day of life. At 4 years of age, a bidirectional Glenn’s operation was performed for the further growth of the pulmonary bed and the volume reduction of the left ventricle. Finally, an extracardiac cavo-pulmonary connection was created using a ringed Gore-Tex graft at 6 years of age. Staged Fontan’s strategy may contribute to a good surgical outcome for this highly lethal anomaly.

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1. Introduction

An unguarded tricuspid orifice is a very rare congenital anomaly. Its main morphological feature is the lack of any guarding valvular tissue at the tricuspid annulus [1—3]. This lesion could be surgically treated by simple tricuspid valve (TV) replacement, but since it is associated with a high incidence of pulmonary valvular abnormalities, successful biventricular repair has rarely been reported. Especially in neonates, unguarded tricuspid orifice with pulmonary atresia (PA) presents with severe hypoxia and right ventricular dysfunction, which results in an extremely high mortality rate. According to earlier surgical reports, no cases have survived except for one that underwent a bidirectional Glenn’s operation [2].

We successfully treated an unguarded tricuspid orifice with PA using a staged Fontan’s operation. To the best of our knowledge, this is the first report of the successful use of this approach.

2. Clinical summary

The patient presented at birth with severe cyanosis and a heart murmur. Two-dimensional echocardiography revealed PA, a small muscular ventricular septal defect, a patent fossa ovalis, and a patent ductus arteriosus. A left Blalock—Taussig shunt was constructed on the 17th day of life. At 4 years of age, he was referred to our unit for further examination and treatment.

Cross-sectional echocardiography demonstrated absence of all tricuspid leaflets, chordae tendineae, and papillary muscles (Fig. 1A). The wall of the RV was thin, and systolic function was severely depressed. Color Doppler examination revealed no tricuspid regurgitation and a to-and-fro pattern with low peak velocity across the tricuspid annulus (Fig. 1B). Cardiac catheterization showed a mean RA pressure of 7 mmHg, a systolic RV pressure of 12 mmHg, and a mean pulmonary arterial pressure of 12 mmHg. Although RV systolic function was severely depressed, LV systolic function was normal. Both the RV and LV were dilated (143% and 336%, respectively). Thus, a bidirectional Glenn’s operation was performed.

At 6 years of age, cardiac catheterization showed a mean pulmonary pressure of 9 mmHg, and the ventricular volumes had decreased (RV, 85% of normal; LV, 221% of normal). A total cavo-pulmonary connection was performed. Inspection of the heart via an atriotomy revealed the smooth surface of the thin anterior RV wall, which had poor contractility. There was no guarding tissue at the dilated tricuspid annulus. No chordae tendineae and papillary muscles were observed in the RV. Although the basal RV muscle had degenerated, trabeculation was observed at the apex (Fig. 1C). An extracardiac cavo-pulmonary connection was created using a ringed Gore-Tex graft. The patient’s postoperative course was uneventful.
3. Discussion

An unguarded tricuspid orifice is a rare congenital anomaly that has a high mortality. It is not simply a TV defect, but is frequently associated with pulmonary valvular abnormalities. This anomaly should be distinguished from similar anomalies, including Ebstein’s anomaly and PA with intact ventricular septum. To make an accurate diagnosis, it is very important to identify the morphological features of the TV using cross-sectional echocardiography and the flow pattern across the TV using color Doppler examination. The main morphological feature is best demonstrated by examining the mural (posterior) leaflet of the TV, which is absent when the orifice is unguarded but downwardly displaced when it is associated with Ebstein’s anomaly [1]. Total absence of tricuspid guarding tissue results in a to-and-fro flow pattern across the tricuspid annulus, which is the characteristic hallmark of an unguarded tricuspid orifice anomaly.

All cases of unguarded tricuspid orifice with PA require surgical intervention during the neonatal period. In such cases, biventricular repair is not feasible due to poor RV function. The combination of hypop RV function and PA, along with an unguarded tricuspid orifice, as was seen in the present case, is considered to be the most lethal variant [1]. We usually treat such anomalies, which require palliative operations, with early bidirectional Glenn’s strategy. Although late referral may have an adverse affect on ventricular dimension and growth of the pulmonary bed, the staged Fontan’s strategy likely contributed to the good surgical outcome in the present case.

Another variant of this anomaly includes a patent RV outflow tract; in these patients, surgical indication would depend on RV function. In most cases, TV replacement is not indicated due to severe RV dysfunction [4]. However, it has been reported that TV replacement was successfully performed in a case with a patent RV outflow tract and good RV function [5].

In summary, we report the first successfully treated case of unguarded tricuspid orifice with PA using Fontan’s operation. The morphological features of this anomaly were preoperatively identified and directly observed during surgery. LV volume reduction and pulmonary bed growth achieved using the staged Fontan’s strategy appear to contribute to a good surgical outcome.

References