Double-Outlet Left Ventricle with Ebstein Anomaly in a Neonate with VACTERL Association
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Double-outlet left ventricle is a very rare congenital cardiac malformation in which both the aorta and the pulmonary artery arise predominantly or exclusively from the morphologic left ventricle. We describe a neonate with esophageal atresia with tracheo-esophageal fistula (so-called VACTERL association) and unusual cardiac anomalies including double-outlet left ventricle, subaortic ventricular septal defect, pulmonary stenosis, and Ebstein anomaly. Many morphologic variations of cardiac malformation have been described in patients with VACTERL association, but to the best of our knowledge, double-outlet left ventricle combined with Ebstein anomaly has never been reported.

Key Words: Cardiac anomaly • Double-outlet of ventricle • Ebstein anomaly • VACTERL association

INTRODUCTION

Double-outlet ventricle is an uncommon cardiac anomaly that only contributes 1% of congenital heart disease. Meanwhile, double-outlet left ventricle (DOLV) accounts for less than 5% in double-outlet ventricle cases. 1-3 Herein, we described a very rare cardiac anomaly of DOLV associated with Ebstein anomaly in a neonate with VACTERL association.

CASE REPORT

A 1-day-old female baby, small for gestational age with birth weight 2.2 kg, was found to have maternal polyhydramnios and decreasing fetal movement during antenatal check up. On examination, she presented decreased activity and bubbling salivation from the mouth. A grade III/VI systolic heart murmur was audible over the left upper sternum. Chest X-ray showed blinded end of esophagus. The patient was finally diagnosed with tracheo-esophageal fistula by computed tomography scan. Echocardiography revealed a septal leaflet of tricuspid valve displaced into the right ventricle cavity, forming an atrialized right ventricle (Figure 1A). There was a D-loop ventricle with a subaortic ventricular septal defect (VSD) and pulmonary stenosis with pressure gradient of 51 mmHg. Both aorta and pulmonary artery originated from the left ventricle, with the aorta right lateral to the pulmonary trunk (Figure 1B). Cardiac catheterization with left ventriculogram confirmed the diagnosis (Figure 2). The patient underwent modified Blalock-Taussig shunt later.

DISCUSSION

Ventriculoarterial connection is considered double-outlet ventricle when both of the great arteries mainly originate from the same ventricle. The occurrence of DOLV is significantly less than that of double-outlet
right ventricle. It is difficult to use embryologic basis to describe the existence of DOLV. Manner et al. tried to explain it by an excessive leftward shift of the embryonic conotruncus, anomalous differential conal growth, or absorption or malorientation of the subarterial portion of the ventricular septum above the crista supraventricularis separating the right ventricle infundibulum from both great arteries. DOLV is often associated with a VSD. The VSD may locate in a subaortic, subpulmonary, doubly committed or remote area. Subaortic VSD is the most common type. Atrioventricular valve anomalies were also noted in 30% of cases, and mostly in the tricuspid valve. Among 109 cases of DOLV reported by Van Praagh et al., only one autopsy case, similar to our case, presented with DOLV, subaortic VSD, pulmonary stenosis and Ebstein anomaly.

Congenital heart disease is the leading combined anomaly of VACTERL association in Scott et al.’s series, with incidence up to 32.1%. The most common heart anomaly is a VSD, which accounted for 22.3%. Cyanotic heart disease was uncommon with only 4.5% in these patients. Yang et al. declared the rate of heart disease association can be as high as 50%. Though congenital heart disease is a common associated anomaly in VACTERL, the combination of DOLV and Ebstein anomaly has never been reported.

REFERENCES


