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CASE REPORT

Transesophageal Echocardiographic Evaluation of Crisscross Heart with Atrioventricular Valve Regurgitation for Fontan Procedure

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ABSTRACT

Crisscross heart (CCH) is a rare congenital cardiac malformation characterized by crossing of systemic and pulmonary venous blood streams at atrioventricular (A-V) level due to an apparent twisting of the heart about its long axis. In the literature, perioperative evaluation of CCH by transesophageal echocardiography (TEE) has not been described. Here we report a rare case of CCH, post Glenn procedure with A-V valve regurgitation, evaluated by TEE for valve repair/replacement followed by completion of Fontan surgery.

Keywords: Atrioventricular valve, Crisscross heart, Double outlet right ventricle, Fontan, Transesophageal echocardiography.

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INTRODUCTION

Crisscross heart (CCH) is a rare congenital cardiac malformation characterized by crossing of systemic and pulmonary venous blood streams at atrioventricular (A-V) level due to an apparent twisting of the heart about its long axis. The frequency of CCH is less than 8 per 10,00,000 live births, and it accounts for <0.1% of congenital heart defects.[1-3]

Crisscross heart is mostly associated with intracardiac defects, the commonest being ventricular septal defect (VSD) with obstruction to pulmonary outflow. Biventricular repair may not be feasible in a proportion of patients. In such cases, the treatment generally includes initial palliation with systemic to pulmonary or cavo-pulmonary shunt followed by completion of Fontan procedure. Although transthoracic echocardiography (TTE) remains the mainstay of the echocardiographic investigation for initial diagnosis, perioperative transesophageal echocardiography (TEE) has a major role in evaluation of cardiac abnormalities and aids in guidance of surgical decisions.

In the literature, perioperative evaluation of CCH by TEE has not been described. Here we report a rare case of CCH, post Glenn procedure, with A-V valve regurgitation, evaluated by TEE for valve repair/replacement followed by completion of Fontan surgery.

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CASE REPORT

A 15-year-old male, weighing 51 kg, presented with history of breathlessness and cyanosis since birth. He had undergone bidirectional Glenn procedure under cardiopulmonary bypass 8 years back with an uneventful postoperative period. Physical examination revealed cyanosis, breathlessness, grade VI clubbing and a mid-sternotomy scar. His heart rate and blood pressure were 100 beats/minute and 100/62 mm Hg respectively. A grade 4/6 systolic murmur was present in third left intercostal space.

The two-dimensional TTE revealed situs solitus, levocardia, superio-inferior arrangement of right and left ventricles, and L posed aorta (aorta on the left and anterior to pulmonary artery). There was a large VSD, with both great arteries originating from right ventricle (RV). The pulmonary valve was thickened, stenosed with gradient of 77 mm Hg. Moderate tricuspid regurgitation was present. The bidirectional Glenn flow was normal. Preoperative cardiac catheterization and angiography showed patent Glenn shunt with good-sized pulmonary artery branches. There was moderate regurgitation of the right A-V valve. The diagnosis of the crisscross position of the ventricles was confirmed.

After securing airway, TEE was performed using X7-2t Philips ultrasound probe and ultrasound machine (Philips iE33 model, Bothell, WA, USA). In midesophageal four-chamber view (Fig. 1) at 0°, instead of all four

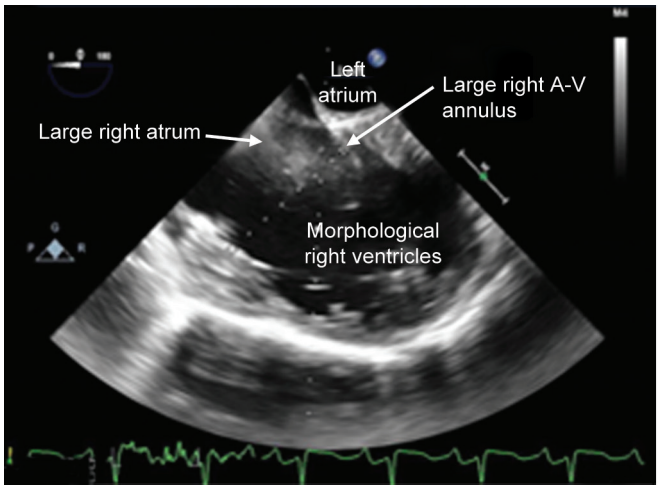

Fig. 1: Midesophageal 0° view, showing large right atrium (confirmed by infusion of saline through right jugular vein), large morphological RV (coarse trabeculations and moderator band), and a large right A-V valve. Unlike in a normal heart, a classical four-chamber view was not visible. AV: Atrioventricular

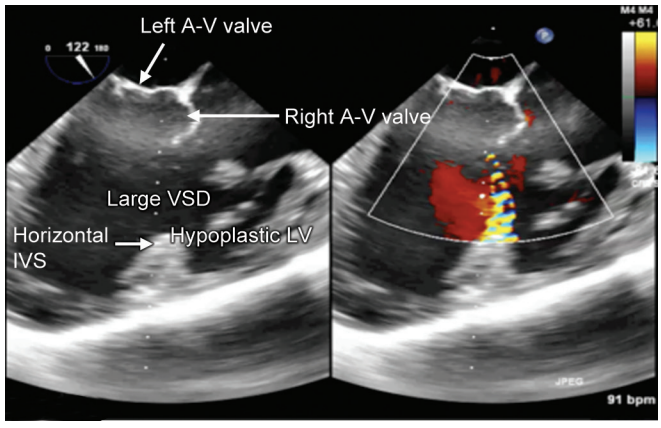

Fig. 2: Midesophageal 120° view, showing all four chambers of heart that are normally seen in midesophageal 0° view. The inflow stream axis of left A-V valve, inlet of LV, and right A-V valve, inlet of RV, are not parallel but angulated to each other. The large RV is receiving blood flow from both left and right atrium (double inlet RV) and LV is hypoplastic. The classical midesophageal aortic long axis view normally seen at 110-130° is not visible. AV: Atrioventricular, VSD: Ventricular septal defect, IVS: Interventricular septum, LV: Left ventricle

chambers, a large right atrial chamber and a large ventricular chamber was visible. A moderate to severe regurgitation was present at the right A-V valve (Video 1). The systemic veins were connected to the right atrium as revealed by rapid infusion of saline through the right jugular venous line. The midesophageal 120° view (Fig. 2, Video 2) revealed all four chambers with a horizontally placed interventricular septum. This view also revealed a large non-restrictive VSD. The left-sided ventricular chamber was hypoplastic and coarse trabeculations with a muscular band was present in the large right-sided ventricle (morphological RV). The two A-V valves were present at the same level, with the right valve significantly larger (5.2 cm) than the left A-V valve (2.4 cm). The inflow stream axis of the left A-V valve and inlet of the left ventricle (LV) were not parallel, but angulated to the axis of the right A-V valve and inlet of the RV. The large RV was receiving blood from both left and right atrium (double inlet RV). In midesophageal 74° view (Fig. 3, Video 3), the aortic and pulmonary valves were visible in the long axis that is usually seen at an angle of 110–130° in normal heart. The two great arteries were originating from the morphological RV, giving a double outlet right ventricle (DORV) appearance (Video 3).

In the bypass period, the surgical evaluation revealed superio-inferior arrangement of right and left ventricles. The aorta was left and anterior to pulmonary artery. The left A-V valve appeared small, and the right A-V valve confirmed moderate to severe regurgitation on saline test. Hence, a decision of right A-V valve replacement with completion of Fontan was taken. The A-V valve was replaced because inadequate repair of the valve

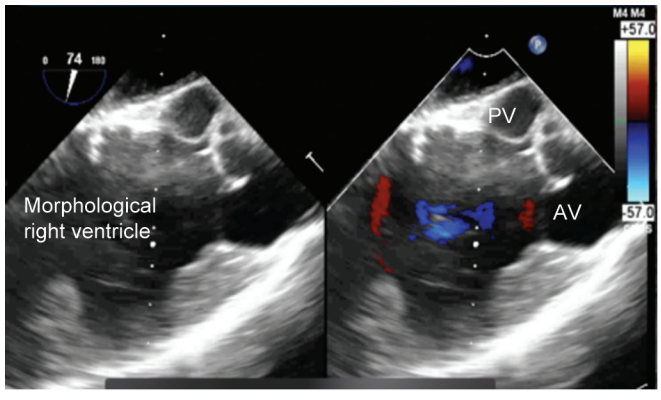

Fig. 3: Midesophageal 74° view, showing both great vessels (aorta and pulmonary artery) originating from right ventricle DORV. Unlike in normal heart, the midesophageal aortic valve long axis view seen at 110–130° was visible at midesophageal 74° view in this patient. PV: Pulmonary valve, AV: Aortic valve

could significantly increase the morbidity after Fontan procedure.

The patient was extubated after 6 hours in intensive care unit and shifted to the ward after 24 hours. The postoperative period was uneventful.

DISCUSSION

Crisscross heart was first described in 1961 by Lev and Rowlatt as an unusual arrangement of the ventricular chambers in a superio-inferior fashion.[2] The term "crisscross" heart was introduced by Anderson et al, for cardiac anomaly producing the illusion of crossing of the pulmonary and systemic venous flow, without mixing at the A-V junction. The etiology of CCH is still not known, but this anomaly seems to be due to abnormal twisting of the apex of the heart, while the base

remains relatively fixed. This accounts for crossing of the systemic and pulmonary venous streams, the abnormal position of the ventricles, and the ventriculo-arterial segmental situs discordance.[3,4] Most of the patients with CCH have associated anomalies, such as VSD, transposition of great vessels, right ventricular hypoplasia, subpulmonary stenosis, and straddling of A-V valves.[5,6]

In two-dimensional TTE examination, failure to obtain a characteristic 4-chamber view is diagnostic for recognition of crisscross A-V junctions. The parallel arrangement of A-V valves and ventricular inlets cannot be achieved and the two valves are not easily seen simultaneously on the apical four-chamber view.[7]

Although TTE is the primary diagnostic tool of congenital heart diseases, TEE helps in perioperative evaluation and decision-making of such complex cardiac anomalies. In this case, patient had undergone bidirectional Glenn procedure 8 years back and staged completion of Fontan procedure was planned. In the pre-cardiopulmonary bypass period, a detailed TEE examination helped in evaluation of A-V valve morphology, annular diameter, A-V connections, and A-V valve regurgitation. In addition, the arrangement of ventricular chambers, function, and ventriculo-arterial relation was also evaluated. In the present case, there was situs solitus, a large right atrium connected to systemic veins and a large right A-V valve with moderate to severe regurgitation. The left A-V valve was small. There was a large nonrestrictive VSD and the left-sided ventricular chamber was hypoplastic, giving univentricular heart morphology. The large right-sided ventricular chamber with coarse trabeculations and a muscular band was connected to both great arteries, giving a DORV appearance.

In the majority of patients with CCH, a two-ventricle repair is not possible because of associated cardiac anomalies. These patients are generally staged toward a Fontan procedure. Mair and colleagues have described the failure of A-V connections as one of the factors contributing to long-term complications after Fontan procedure.[8] Podzolkov et al also found that, if unrepaired, concomitant moderate to severe A-V valve regurgitation significantly worsens the results of Fontan procedure.[9] In the present case after TEE examination, a surgical decision of right A-V valve replacement was taken as the inadequate repair of valve could significantly increase the morbidity after Fontan procedure. So the replacement of right A-V valve along with completion of Fontan procedure was done.

In the literature, TTE and angiographic study findings of CCH have been mentioned earlier,[10,11] but perioperative TEE evaluation has not been described. In this case, we observed that midesophageal TEE views were very different from classic TEE views of a normal heart.

To conclude, a detailed TEE examination in the perioperative period can help the surgeon to analyze such complex cardiac anomalies and help decide the surgical plan for a better patient outcome.

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