Case series: Criss-cross Heart

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Abstract

Criss-cross heart (CCH) is a rare cardiac malformation which is characterized by crossing of the inflow streams of the two ventricles due to rotation of ventricular axis. The anomalies can be identified both atrioventricular concordance and discordance. The etiology of CCH is remaining unknown. Prominent symptom is cyanosis. The primary investigation is transthoracic echocardiography to identify this abnormality. Many patients need further investigation to review anatomy. Total correction is the aim for treatment but it is difficult for this complex anatomy. There were 5 patients who visited cardiology clinic at Queen Sirikit National Institute of Child Health (QSNICH) from 2002 to 2017. The objective is to review the treatment options that we performed in CCH. Most common associated anomalies were double outlet right ventricle and pulmonary stenosis. One patient died before surgical intervention; others were performed palliative surgery.

Introduction

CCH is a rare congenital heart defects accounting less than 0.1% of all congenital heart disease, characterized by crossing of the inflow stream of the two ventricles due to rotation of ventricular axis as shown in figure 1 [1]. The etiology of CCH is remaining unknown. Other anomalies such as ventricular septal defect (VSD), double outlet right ventricle (DORV), dextrocardia, overriding atrioventricular (AV) valve, pulmonary stenosis, pulmonary atresia or straddling atrioventricular (AV) valve are seen [2–4]

The diagnosis requires transthoracic echocardiography for initial diagnosis [5, 6]. In case of further information needed, additional examination may be performed to help diagnose cardiac anatomy for treatment planning such as 3-Dimensional echocardiography, transesophageal echocardiography, cardiac intervention, or computed tomography angiography [7–8]. There are two types of operations to treat criss-cross heart: biventricular repair and univentricular repair. A surgical complexity depends on an individual's cardiac anatomy.

At present, there are many criss-cross heart cases at QSNICH which are treated according to associated anomalies. However, there are no data collection of disease development, comorbidity and treatment results.

Case 1

An 18-day-old male infant with cyanosis admitted at QSNICH and was found to have pansystolic murmur, grade III at left parasternal border, no hepatomegaly, and oxygen saturation was 68%. Chest radiography showed normal cardiothoracic ratio, decreasing of blood flows to the lungs, and normal EKG. Echocardiogram then showed situs solitus, criss-cross heart, DORV, VSD, and pulmonary stenosis. He underwent right modified Blalock Taussig shunt (RMBTS) and left modified Blalock Taussig shunt (LMBTS) at 11 months. At the age of 4 years and 4 months, he developed severe cyanosis and got better after receiving revised RMBTS and atrial septectomy procedure. Cardiac computed tomography (CT) was

performed when he was 6 years old. The image showed that shunt was patent, small pulmonic annulus, and low McGoon ratio (1.25). However, cardiac catheterization was to evaluate hemodynamics and demonstrated high pulmonary arterial pressure (Mean 20 mmHg) and left ventricular end diastolic pressure (15 mmHg). He currently has an appointment for a follow-up at Cardiology Clinic with 83% of SpO₂.

Case 2

A 19-year-old male was referred to QSNICH. He presented with cyanosis and underwent RMBTS at 7 years old. His echocardiogram revealed situs solitus, dextrocardia, criss-cross heart, DORV, large VSD, large ASD, and pulmonary stenosis. RMBTS was done when he was 6 years old. He was doing well until 17 years of age, he had worsening cyanosis then imaging were evaluated. Cardiac CT showed patent RMBTS and could not repair VSD then diagnostic catheterization confirmed good hemodynamics for univentricular repair. He underwent median sternotomy, extracardiac Fontan with Gortex No.22, 5 millimeters of fenestration, main pulmonary artery (MPA) interruption, right pulmonary artery plasty, taken down RMBTS and patent ductus arteriosus (PDA) closure. His saturation was improved after operation. He still visits Cardiology Clinic for follows up appointment from time to time.

Case 3

A 15-year-old female with cyanosis was referred to QSNICH. Her chest film showed decreased pulmonary vasculature. Echocardiogram showed situs solitus, levocardia, criss-cross heart, and pulmonary atresia. Diagnostic catheterization confirmed good hemodynamics for univentricular repair when she was 16 years old. Then she underwent median sternotomy, glenn shunt with Gortex No.22, and pulmonary artery reconstruction, left pulmonary artery enlargement, atrial septectomy, taken down RMBTS and PDA closure when she was 16 years and 6 months old. Permanent pacemaker was implanted after surgery, her condition was improved and regular follow up with cardiology clinic.

Case 4

A 1-year-old male presented with cyanosis. Cardiac auscultation showed systolic ejection murmur grade III/VI left parasternal border, no hepatomegaly. Chest film showed normal cardiothoracic ratio and decreasing of blood flows to the lungs. EKG showed right atrium enlargement, and right ventricular hypertrophy. Echocardiogram showed situs solitus, dextrocardia, criss-cross heart, DORV, VSD, and pulmonary stenosis. Figure 1 and 2 were transthoracic echocardiogram of criss-cross heart and DORV. Median sternotomy, Hemi-Fontan, MPA interruption, atrial septectomy, and PDA division were performed when she was 3 years old. His saturation has improved after the procedures and continued her appointment at cardiology clinic until present.

Case 5

A 9-month-old female baby visited to the hospital and presented with cyanosis. Cardiac auscultation showed systolic ejection murmur grade III/VI left parasternal border, no hepatomegaly. Chest film showed normal cardiothoracic ratio and decreasing of blood flows to the lungs. EKG was normal. Echocardiogram revealed situs inversus, dextrocardia, criss-cross heart, DORV, VSD, pulmonary stenosis and anomalous origins of coronary artery. Cardiac CT shows unrepairable VSD and cardiac catheterization was scheduled. However, she did not undergo any operations and died at the age of 9 with unknown cause.

Discussion

Criss-cross heart report was first described in 1974, which was characterized by crossing of the inflow streams of the two ventricles due to rotation of ventricular axis. The anomalies identify both atrioventricular concordance and discordance. Echocardiogram helps to diagnosis and identify this abnormality. The symptom of patients usually depends on associated cardiac anomalies. Anderson RH et al. stated that criss-cross heart could be identified when the anatomically RA is connected to the anatomically RV, and the anatomically LA to the anatomically LV (atrioventricular concordance) or when the anatomically RA is connected to the anatomically LV & the anatomically LA to the anatomically RV (atrioventricular discordance) [9]. Biventricular repair preferred than univentricular repair for criss-cross heart.

Junichi Kashiwagi et al. reported that biventricular repair was performed to treat criss-cross heart. Total surgical correction could effectively treat patients with criss-cross heart associated with VSD, transposition of great vessel, and PDA [10]. This study has included patients at QSNICH for 14 years and found total 5 cases. Criss-cross heart associated with other cardiac malformations such as DORV (5 patients), pulmonary stenosis (4 patients), dextrocardia (3 patients) and pulmonary atresia (1 patient). DORV is most common associated with criss-cross heart. This study reported that 4 of 5 DORV patients underwent a particular type of operations: palliative modified Blalock Taussig shunt, Glen shunt, Extracardiac Fontan and Hemi-Fontan. One patient died before receiving surgery.

In conclusion, biventricular repair could not be performed in all 4 patients, but they all clinical condition was improving after surgery. 3D printing might be effective technologies for treatment preparation and planning in the future.

Conclusion

The treatment of criss-cross heart need to assess cardiac anatomy thoroughly by pediatric cardiologists, cardiothoracic surgeons, and radiologists to plan the best suitable operation for each patient.

Declarations

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Ethic statement

- There is no conflict of interest.
- The study has been approved by the Queen Sirikit National Institute of Child Health ethics committee (Number REC.039/2563) and has been performed in accordance with the ethical standards as laid down in the 1964 Declaration of Helsinki and its later amendments or comparable ethical standards.
- Informed consent was obtained from all individual participants included in the study.

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Figures

Figure 1

A: Circulatory system of atrioventricular concordance, which right atrium (RA) connected to a right ventricle (RV) and left atrium (LA) connected to a left ventricle (LV).

B: Circulatory system of atrioventricular discordance, which right atrium (RA) connected to a left ventricle (LV) and left atrium (LA) connected to a right ventricle (RV)

Figure 2

Subcostal view of TTE shows left ventricle (LV) located on the bottom right. Right ventricle (RV) located on the top left connected to aorta.

Figure 3

Subcostal view of TTE shows left ventricle (LV) located on the bottom right.

Right ventricle (RV) located on the top left connected to narrowed pulmonary artery.