Rastelli Procedure in Tetralogy of Fallot Patient with Anomalous Coronary Artery: A Case Report

Achmad Aulia Rachman(✉), Heroe Soebroto, Arief Rakhman Hakim, and Erdyanto Akbar

Department of Cardiac, Thoracic, and Vascular Surgery, Dr Soetomo General Hospital, Universitas Airlangga, Surabaya, Indonesia
achmad.aulia.r@gmail.com

Abstract. Tetralogy of Fallot (ToF) is the most common form of cyanotic heart disease with a prevalence between 3,5 to 8%. Anomalous coronary artery is reported in 2–23% of ToF patient. Knowledge of the coronary anatomy prior to surgery was important to avoid injury to the vessel. In our case, ToF with the presence of coronary artery that crossing the right ventricle outflow (RVOT) tract altered our strategy of surgery using right ventricle-pulmonary artery bypass (Rastelli procedure) in order to avoid injury to the vessel. Case report: A 2-years-old boy was presented with repeated respiratory complaint and delayed both in growth and development. Cyanosis and clubbing fingers were observed in this patient. Overriding aorta and infundibular-valvular pulmonary stenosis were found on transthoracic echocardiography exam and additional finding was right coronary artery crossing the RVOT. The patient underwent total correction of ToF with additional procedure of the right ventricle to pulmonary artery bypass (Rastelli Procedure). The surgery was successful and the length of stay of the patient was eight days. One month following the surgery, we evaluated the flow in the conduit was preferable. The case represented the possible alternative management for ToF with anomalous coronary artery.

Keywords: tetralogy of fallot · Rastelli procedure

1 Introduction

The incidence of congenital heart disease was 134/10.000 person-years. Tetralogy of Fallot (ToF) was 3,5–8% among them. Anomalous coronary artery was more common in ToF patients than in general population. Some anomalies might have significance in the disease, such as coronary artery crossing the right ventricular outflow tract (RVOT).[1] In our case, the presence of vessels that cross the RVOT altered our strategy of surgery.

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2 Case

A 2 years old boy (weight: 10 kg, height: 90 cm) was presented to us with repeated respiratory complaint and delayed growth and development. History of shortness of breath and squatting was reported by his parent. Cyanosis and clubbing finger were observed in this patient. His vital sign was as follows: Peripheral SpO2 of 75%, heart rate of 95 beats per minute, respiratory rate of 26 per minute. Systolic murmur was heard on auscultation on the second and fourth left parasternal line. Chest x-ray showed right ventricle hypertrophy with upward apex and diminished pulmonary vascular pattern (Fig. 1).

Trans-thoracal echocardiography showed malalignment ventricular septal defect (diameter of 1,53 cm), overriding aorta (Fig. 2), and infundibular-valvular pulmonary stenosis with pressure gradient of 57,90 mmHg (Fig. 3). Additional finding was single ostium left coronary artery (Fig. 4) which crossed the RVOT. The diameter of left and right pulmonary artery were 1,03 cm and 1,15 cm respectively.

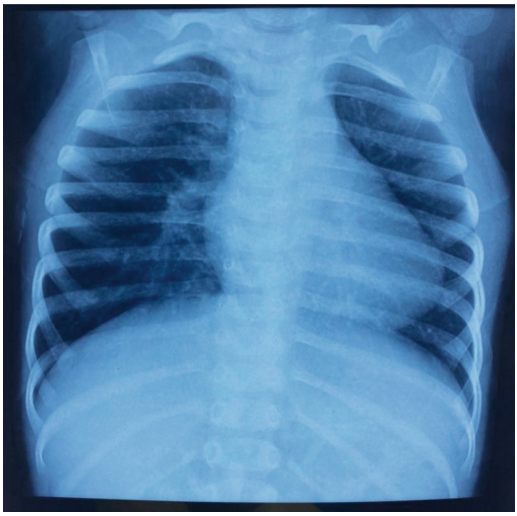

Fig. 1. Chest X-ray showed right ventricular hypertrophy and diminished pulmonary vascular pattern

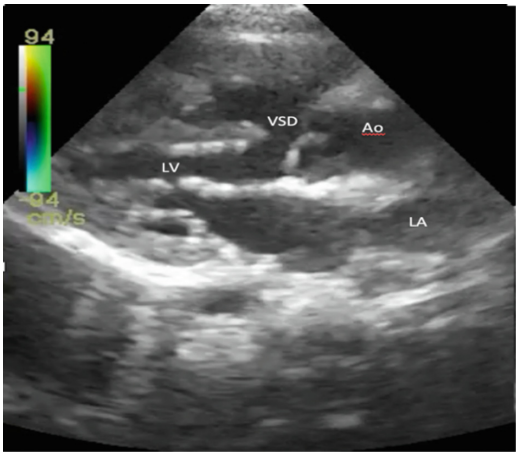

Fig. 2. Parasternal long axis view showed malalignment VSD with overriding aorta.(LV: Left ventricle; LA: left atrium; Ao: aorta; VSD: ventricular septal defect)

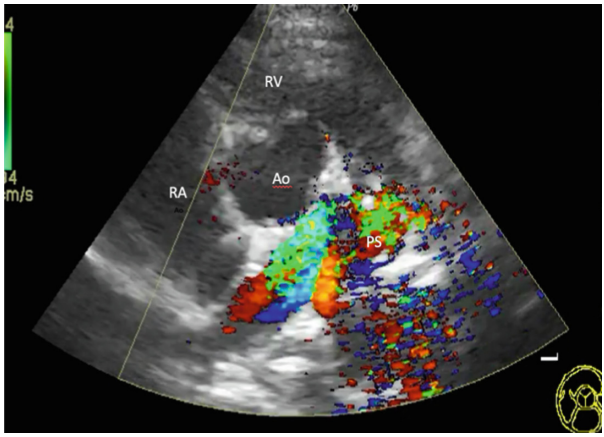

Fig. 3. Parasternal short axis view showed jet of the pulmonary stenosis. (RA: right atrium; RV right ventricle; Ao: aorta; PS: pulmonary stenosis)

The patient underwent total correction of ToF with additional right ventricle to pulmonary artery bypass (Rastelli Procedure). At first, patent ductus arteriosus was identified, it was ligated and divided. Left main coronary artery was found crossing the RVOT (Fig. 5). We use trans-atrial approach to close the VSD with polytetrafluoroethylene patch. Right ventriculotomy was done, and ventricular muscle was resected to increase the diameter of the right ventriculotomy. The proximal main pulmonary artery (MPA) was ligated. We use bovine jugular vein conduit to bypass the flow from the right ventricle (RV) to pulmonary artery (Fig. 6). End to side anastomosis from the distal conduit to the MPA (subsequent the ligation site) was done first. Proximal conduit was anastomosed to the RV. The duration of cardiopulmonary bypass machine time was 170 min

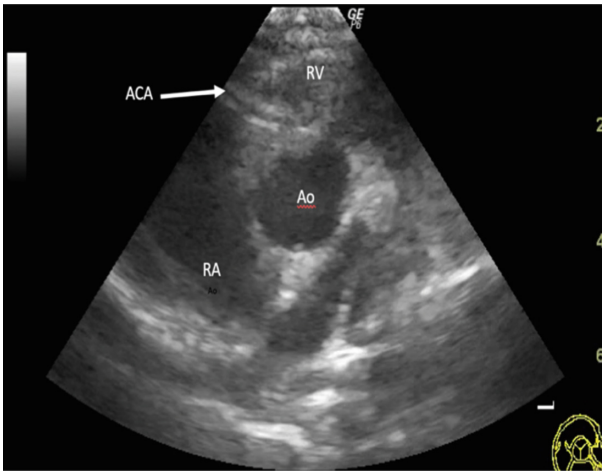

Fig. 4. Parasternal short axis showed anomalous coronary artery crossing the RV. (ACA: anomalous coronary artery)

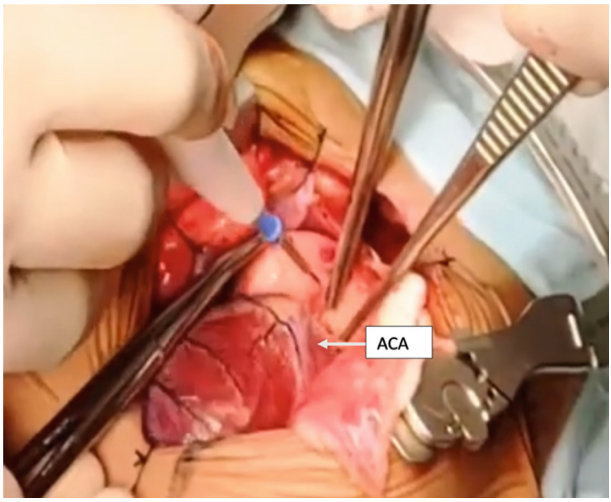

Fig. 5. Left main coronary artery was found crossing the RVOT

and the aortic cross clamp time was 134 min. Trans-epicardia echocardiography evaluation showed adequate flow on the RV-PA conduit, and acceptable residual flow on the VSD. He was observed in intensive care for three days, and discharged from the hospital eight days following the surgery. One month following the surgery, we evaluated the flow in the conduit was preferable.

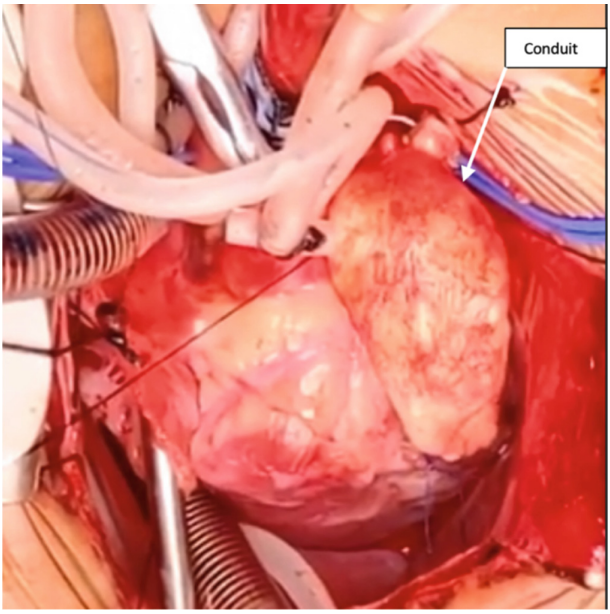

Fig. 6. Bovine jugular conduit was used to connect right ventricle to the pulmonary artery

3 Result and Discussion

Among person with congenital heart defect, 5% of them is ToF, which consists of ventricular septal defect, overriding aorta, pulmonary stenosis and right ventricular hypertrophy. Anomalous coronary artery (ACA) is also frequently found in ToF patient than in the general population. However, not all ACA shows significant clinical effect. One of the ACA that significantly shows clinical effect is vessels which cross the RVOT. ACA crossing the RVOT increase the risk of coronary artery injury that may lead to myocardial infarction following the total correction procedure. This abnormality can be detected prior to surgery by using echocardiography or angiography.[1].

Our case showed left main coronary artery crossed the RVOT. ACA on ToF case may vary to some types of abnormality, such as left anterior descending (LAD) artery which come from the right coronary artery (RCA), single RCA ostium, RCA coming from the LAD, single left coronary (LCA) ostium, ACA crossing the RVOT, and other anomalies.[2].

We used bovine jugular vein graft to bypass the blood flow from RV to PA in order to avoid injury to the coronary artery. Options for repair techniques in the setting of major coronary artery crossing the RVOT are repair approach trough the RA and pulmonary artery (Rastelli procedure), RV to PA bypass using conduit, and limited right ventriculotomy with or without mobilization of the coronary artery.[3].

In our case the procedure was successful and the patient was discharged with excellent result. Rastelli procedure may be a safe option for case such as ToF with major coronary artery crossing the RVOT. The early mortality rate of the Rastelli procedure is considered to be around 5%. Most of the patient will require reoperation for adjusting the size of

the conduit later when the child is growing older, but this procedure is reported with low morbidity and mortality. One study also shows excellent outcome of Rastelli procedure done on patients with RVOT obstruction.[4].

4 Conclusion

Rastelli procedure can be a safe surgical approach for repair of ToF with ACA crossing the RVOT in order to avoid injury to the vessel.

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Author's Contribution. AAR, HS, ARH, EA managed and treated the patient, wrote and organized the manuscript.

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