

TRUNCUS ARTERIOSUS TYPE I: A CASE REPORT

Putu Fristia Indarini¹, Eka Gunawijaya²

¹*Department of Child Health Udayana University, Sanglah Hospital*

²*Medical School of Udayana University, Sanglah Hospital*

Corresponding: Fristia, e-mail: fristiaindarini@me.com

ABSTRACT

Truncus Arteriosus (TA) is a rare congenital cyanotic heart disease which is characterized by single arterial trunk arising from heart which in turn gives rise to pulmonary trunk in various ways. TA constitutes 1.2% of all congenital heart malformations. According to Collett-Edwards classification there are 4 types of the disease. The most common variety is Type-1 which is characterized by short main pulmonary artery that originate from the trunk and give rise to right and left pulmonary arteries. Early surgical intervention is advised in all cases within first 2 month of life, in case of severe pulmonary hypertension, surgery is contraindicated. A full-term male newborn suspected with congenital cyanotic heart disease is referred to Sanglah Hospital. The patient had respiratory distress and looked cyanotic after breastfed. Physical examination revealed murmur on the chest auscultation. The chest X-ray showed prominent pulmonary vascular markings and unusually high hilar areas. The echocardiography showed truncus arteriosus type I, with moderate regurgitation, large pulmonary atresia, large ventricular septal defect, functionally single atrium, moderate mitral regurgitation, mild tricuspid regurgitation. Patient was also diagnosed with heart failure ROSS II. The patient was given milk through orogastric tube, oxygen, and oral therapy for heart failure such as furosemide, spironolactone and digoxin. Another echocardiography was planned in the next 3 month. Pulmonary hypertension, heart failure, infective endocarditis and desaturation are recommended to be closely monitored.

Keywords: truncus arteriosus, heart failure

INTRODUCTION

Truncus arteriosus (TA) constitutes 1.2% of all congenital heart malformations.¹ In TA, systemic pulmonary and coronary circulation originate from a single vessel that is emerging from the right and left ventricles by a truncal valve usually tricuspid, but quadricuspid and bicuspid valves have also been reported.² The valve leaflets may be up to six in number.³ This condition may lead to valvular regurgitation, stenosis or both. Patients with TA typically have a large subarterial ventricular septal defect (VSD) located beneath the truncal valve.

TA classification is made by two systems developed by Collett-Edwards and Van Praagh, which are based on the origins of the pulmonary

arteries.^{4,5} According to Collett-Edwards classification, type 1 disease is characterized by a single pulmonary arterial trunk that originates from the common trunk and gives off right and left pulmonary arteries. While in Type 2 TA, main pulmonary arterial trunk is absent and the right and left pulmonary arteries originate together. In type 3 TA, there is no pulmonary truncus present, however, the right and left pulmonary arteries branched from the truncus separately. Type 4 is identified by the absence of pulmonary arteries; the lungs are supplied by major aorto-pulmonary collateral arteries, which form an anatomic picture that may also be named as pulmonary atresia with VSD.

In Van Praagh classification system, TA Type A1 and A2 are similar to Collet-Edwards TA Type 1 and Type 2, respectively. Type A3 is characterized by a branch of pulmonary artery (usually right) originating from the common trunk while other lung is perfused by either the pulmonary artery originating from the aortic arch or a vessel with an origin from ductus arteriosus or major aorto-pulmonary collaterals. There is an associated interrupted aortic arch in addition to Type 1 and 2 in Type A4.

The coronaries usually originate independently and have a normal course. However, rare anomalous coronary take offs or courses may also be observed. 6% of cases are characterized by a persistent left superior vena cava (PLSVC) draining into coronary sinus.³ The disorder is accompanied by an interrupted aortic arch or an aortic coarctation in 14% of cases. A patent foramen ovale and an atrial septal defect are common. However, a coarctation of the aorta, an anomalous pulmonary venous drainage, a hypoplasia of the left lung, an anomalous origin of subclavian arteries, a single ventricle, an absent right arteries-veins connection, a hypoplastic right ventricle, a tubular hypoplasia of the aortic arch, and a double aortic anomaly may rarely present.³

CASE REPORT

An eighteen-days-old male baby was referred from a private hospital in West Nusa Tenggara, to our Sanglah Hospital's emergency room, Bali, with main complain respiratory distress and cyanosis. Based on the echocardiography, he was suspected with a transposition of the great artery (TGA), a large peri-membrane ventricular septal defect (VSD) and a single functional atrium.

The patient was the fourth child, with no history of congenital heart defect in both parents nor his second grade family. He was born as a vigorous baby, with a body weight of 4100 grams, a body length of 52 cm. There was no illness during the pregnancy period noted, nor a history of a traditional medicine consumption. The patient was delivered at full term gestation with a cesarean section due to an early membrane rupture.

Antenatal care was performed by an obstetrician regularly and the antenatal ultrasound was normal. Vitamin K and hepatitis B immunization was performed on the first hour of life.

On the 10th day of life, the patient looked cyanotic and unchanged by a positional change. The parents also complaint about the poor feeding. Then, he was then brought to a private hospital in which a chest x-ray (**Figure 1**) and an echocardiography were performed. The chest X-ray showed a prominent pulmonary vascular markings and an unusually high hilar areas. The echocardiography, performed by a general pediatrician, showed a suspected TGA, a large perimembrane VSD and a single functional atrium. The doctor and family decided to refer this patient to Sanglah hospital for further examination.

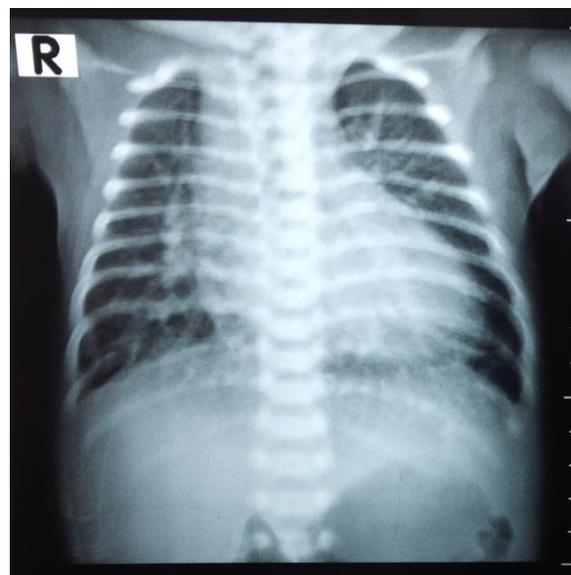


Figure 1. The chest X-ray

He was admitted to our hospital on August 31, 2014. A physical examination revealed an alert baby, a heart rate of 140 times per minute, regular, and the respiration rate was 60 times per minute, regular. The peripheral oxygen saturation was 86%. The body temperature was 36.7°C. Body weight on admission was 4300 gram. The head was normal-shape and the hair was thick, black, and the fontanel was flat. There was no jaundice on sclera, neither conjunctiva injection, anemia, nor sunken. The pupil

light reflex was normal. The ear, nose, and throat examination were within normal limit. The sucking reflex was normal. There was no lymph node enlargement on the neck. The chest was symmetrical both on rest and during respiration. On auscultation, the breath sound was bronchovesicular without rales or wheezing. The first and second heart sound were normal, regular and murmur is audible on the fifth left intercostal space (ICS), grade III/6. There was no lymph node enlargement on both of the axillas

The skin elasticity was normal, no epigastric distention, the bowel sound was normal, the liver was palpable about 2 cm below costal arc and 2 cm below xiphoid process and the spleen were not palpable. There was no inguinal lymph node enlargement found on both sides. The extremities were normal. Moreover, the power, tonus, and the reflexes of the superior and inferior extremities were normal.

The patient was diagnosed with a congenital heart defect: a suspected TGA, a large perimembrane VSD, a single functional atrium and a heart failure ROSS II. He was given a breastmilk on demand via orogastric tube, oxygen, and an oral therapy consists of furosemide and spironolactone. Echocardiography was planned for this patient.

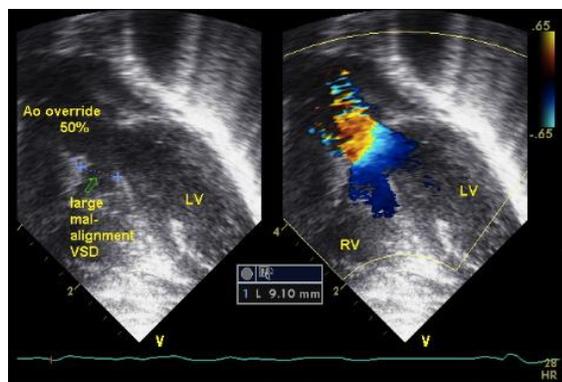


Figure 2. The echocardiography showed a VSD

The second echocardiography, performed by a pediatric cardiologist, showed truncus arteriosus type I, with moderate regurgitation, large pulmonary artery (PA), large ventricular septal defect, single functional atrium, moderate mitral regurgitation and

mild tricuspid regurgitation (**Figure 2, 3**). Based on the clinical manifestation, the chest x-ray, and the echocardiography, the patient was diagnosed with a heart failure ROSS II, truncus arteriosus type I, with moderate regurgitation, large PA, large VSD, functionally single atrium, moderate MR, mild TR. Digoxin was added as an oral therapy. The echocardiography will be repeated in 3 months. A close monitoring of the early signs of a pulmonary hypertension, a heart failure, an infective endocarditis and a desaturation, was performed. A surgical intervention was not performed because such procedure cannot be done in our medical center.

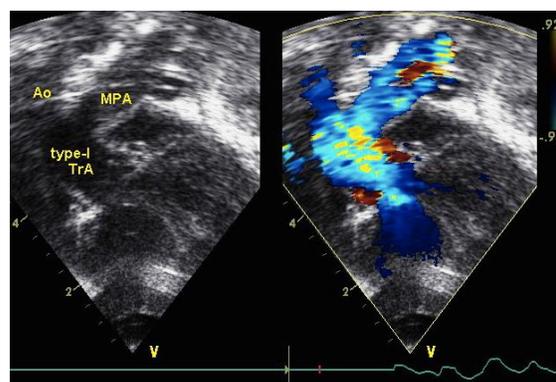


Figure 3. The echocardiography showed truncus arteriosus type I

On September 9, 2014, this patient was transferred to the Mataram Hospital in his home province, West Nusa Tenggara, to continue his treatment.

DISCUSSION

Truncus arteriosus is a rare congenital cyanotic heart disease characterized by a single arterial trunk arising from the heart, which gives rise to pulmonary trunk. There are four types of pulmonary trunk recognized, the most common variety is type 1, characterized by short main pulmonary artery that originate from truncus and give rise to right and left Pulmonary arteries. In type 2, the pulmonary artery arises from separate ostia from the side of truncus. In type-3, the pulmonary arise from a separate ostium from the back of the truncus. In type 4, the pulmonary artery is absent or

atretic. The truncal valve could be quadricuspid (40-50%), bicuspid, tricuspid or hexacuspid. The left coronary artery arise from the left posterior aspect of the truncus, the right coronary artery arise from the right anterior aspect of the truncus. A VSD is always nonrestrictive, and roofed by a truncal valve, setting a stage for truncal valve regurgitation.^{1,2} In our patient, the echocardiography showed truncus arteriosus type I, with moderate regurgitation, a large pulmonal artery, a large ventricular septal defect, a single functional atrium, moderate mitral regurgitation, mild tricuspid regurgitation.

A case of truncus arteriosus usually shows a tachypnea, a poor feeding, and a sign of failure to thrive within the first few weeks of life. The patients seldom reach their first birthday.³ Our patient was brought to the hospital on the 10th day of life with a main complaints of a respiratory distress, a cyanosis,

and a poor feeding. His birthweight was 4100 grams, and only gain 200 grams after 18 days of life.

On examination, there are evidence of a cardiomegaly, a collapsing pulse, a loud single second heart sound, a systolic murmur (harsh, blowing, 3/6 – 4/6, best heard on the 3/4 left intercostal space), preceded by an ejection click. Initially, the pulmonary vascular resistance was low. As the pulmonary ventricular resistance (PVR) rises after birth, the pulmonary vascular resistance becomes equal to systemic and causes Eisenmenger syndrome, cyanosis, and bidirectional shunting.^{4,5} The patient was cyanotic and a grade III/6 murmur was audible on the fifth left intercostal space. The liver was palpable about 2 cm below the costal arc and 2 cm below xiphoid process, as signs of a heart failure.

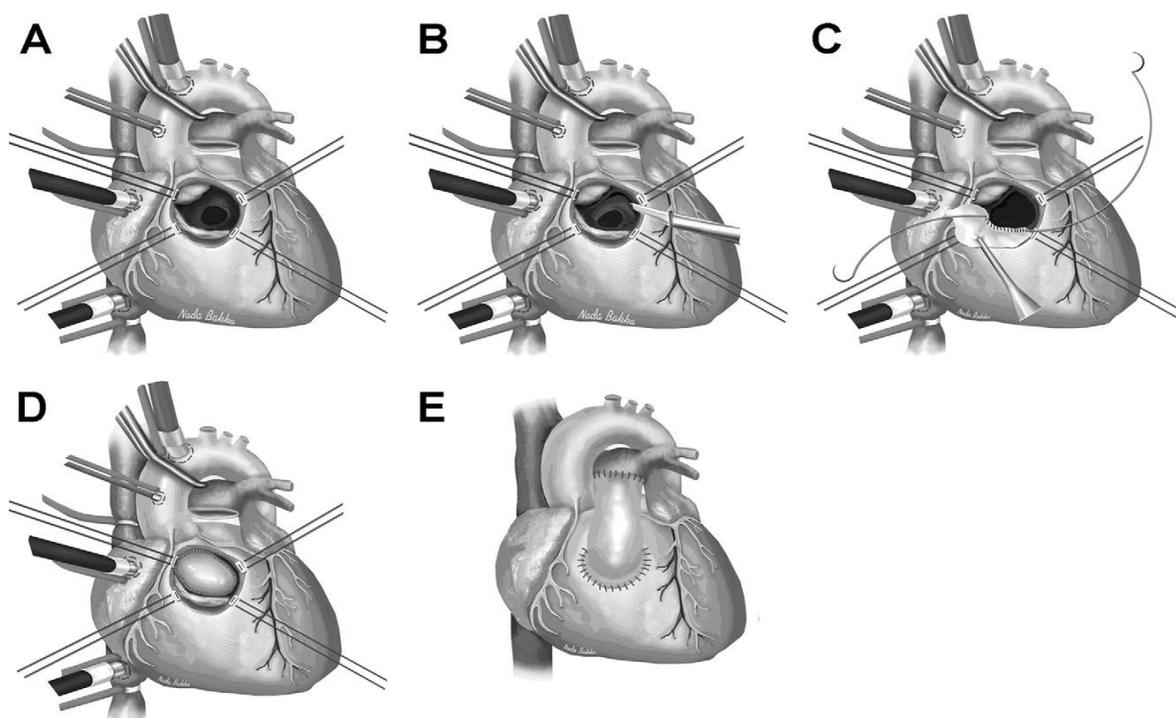


Figure 4. The surgical method used in the Rastelli cohort

(A) Right ventricle incision is placed away from epicardial vessels and retraction sutures are placed to provide exposure to the VSD and tricuspid valve apparatus. **(B)** The VSD is routinely enlarged by excision of a wedge of the septal tissue anterosuperiorly using an 11 blade. In addition, excision of a large portion of infundibular septum beneath the aortic valve is usually done. **(C)** A cylindrical patch is used to construct the baffle with running sutures to the right ventricular tissue. **(D)** The patch should be redundant to provide an unobstructed baffle between the left ventricle and aorta. **(E)** Right ventricle outflow reconstruction is completed with a homograft valved conduit.

An ECG should show a biventricular hypertrophy, and a chest X- Ray should show a cardiomegaly with a prominent pulmonary vascular markings and an unusually high hilar area.^{6,7} However, an ECG was not performed due to the unavailability of ECG probes for neonates. But, the chest x-ray showed a confirmation of the expected.

An early surgical intervention is advised in all cases within the first 2 month of life. In case of a severe pulmonary hypertension, surgery is contraindicated.^{8,9} Operation consist of closure of VSD, leaving the aorta arising from left ventricle; excision of pulmonary arteries from their truncus origin; and a valve containing prosthetic conduit or aortic homograft valve conduit are inserted between the right ventricle and pulmonary arteries to establish circulatory continuity (Rastelli procedure).^{10,11,12} A careful follow up for pulmonary hypertension and truncal conduit patency should be done regularly. An endocarditis prophylaxis is required in all patients. Due the limitation of our medical center, surgical intervention cannot be performed. Therefore, he was given oxygen, and an oral therapy for heart failure: furosemide, spironolactone and digoxin. Another echocardiography will be conducted after 3 months, with a careful monitoring for pulmonary hypertension, heart failure, infective endocarditis and desaturation.

SUMMARY

A full-term male newborn, referred to Sanglah Hospital due to congenital heart defect suspicion. The case was confirmed as a case of TA type type I, with moderate regurgitation, a large PA, a large VSD, and a single functional atrium, moderate MR, and mild TR. He was given an oral medication of furosemide, spironolactone and digoxin, and was planned for an echocardiography in 3 months. A careful monitoring for pulmonary hypertension, heart failure, infective endocarditis and desaturation were commended.

REFERENCES

1. Slavik Z, Keeton BR, Salmon AP, Sutherland GR, Fong LV, Monroe JL. Persistent truncus

- arteriosus operated during infancy: Long-term follow-up. *Pediatr Cardiol* 1994;15:112-5.
2. Crupi G, Macartney FJ, Anderson RH. Persistent truncus arteriosus. A study of 66 autopsy cases with special reference to definition and morphogenesis. *Am J Cardiol* 1977;40:569-78.
3. Williams JM, de Leeuw M, Black MD, Freedom RM, Williams WG, McCrindle BW. Factors associated with outcome of persistent truncus arteriosus. *J Am Coll Cardiol* 1999;34:545-53.
4. Collett RW, Edwards JE. Persistent truncus arteriosus; a classification according to anatomic types. *Surg Clin North Am* 1949; 29:1245-70.
5. Van Praagh R. Truncus arteriosus: what is it really and how should it be classified? *Eur J Cardiothorac Surg* 1987; 1:65-70.
6. Kodo K Nishizaway, Furutani M Yamamure E, Joo K et al GATA6 Mutation causes human cardiac outflow tract defect by disrupting semaphorin-plexin signaling. *Proc Natl Acad Sci USA* Aug 18 2009;106(33):13933-8.
7. Lund AM, Vogel M, Marshal AC, Emani Sm, Pigula FA, Tworetzky W, et al. Early intervention on the pulmonary artery and Right ventricular outflow tract after neonatal or early infant repair Of Truncus Arteriosus using homograft conduits, *American journal of Cardiology* Jul1 2011;108(1):106-13.
8. Alexiou C, Keeton BR, Salmon AP, Monroe JL. Repair of Truncus Arteriosus in early infancy with antibiotic sterilized aortic homografts. *Ann Thoracic surg.* May 2001;71(5 supplement):371-4.
9. Swanson TM, Selamet Tierney ES, Tworetzky W, Pigula F, Mc Elhinney DB. Truncus Arteriosus: diagnostic accuracy, outcome and impact of prenatal diagnosis. *Pediatr Cardiol.* Apr 2009; 30(3):256-61.
10. Russel HM, Pasquali SK, Jacobs ML, O'Brien SM, Mavroudis C et al. Outcome of repair of common arterial trunk with truncal valve surgery: a review of the society of Thoracic surgeons congenital Heart surgery data base.

- Am Thorac Surg. Jan 2012; (1) :164-9.
11. Chen JM, Glickstein JS, Davies RR, et al. The effect of technique on post operative right sided obstruction in patient with Truncus Arteriosus. J Thorac Cardiovascular Surg. March 2005;129(3):559-68.
 12. Alsoufi B, Abid A, Ahmad AO, Mamdouh AA, Charles CC, Ziad B, Avedis K, Zohair AH. The Rastelli Procedure for Transposition of the

Great Arteries: Resection of the Infundibular Septum Diminishes Recurrent Left Ventricular Outflow Tract Obstruction Risk. Ann Thorac Surg. 2009; 88:137-43.

