Jurnal Healthsains
Printed ISSN: 2722-7782
Electronic ISSN:2722-5356
jurnal.healthsains.co.id

BIDIRECTIONAL CAVOPULMONARY SHUNT SURGERY OF TETRALOGY OF FALLOT WITH SITUS INVERSUS AND LEVOCARDIA IN 20-MONTH-OLD BOY :A RARE CASE REPORT

Suprayitno Wardoyo, Dhama Shinta Susanti, William Makdinata, Albert Jeo
Division of Cardiothoracic and Vascular Surgery, Department of Surgery, Faculty of Medicine, Universitas Indonesia, Cipto Mangunkusumo General Hospital,Indonesia
General Practitioner, Universitas Methodist Indonesia
Email: albertjeo@gmail.com

Keywords:
Bidirectional Cavopulmonary Shunt, Congenital Heart Disease, Levocardia, Situs Inversus, Tetralogy of Fallot

ABSTRACT
Tetralogy of Fallot (TOF) is a relatively common cyanotic congenital heart disease, characterized by four distinct defects. Although TOF is a common condition, its association with situs inversus and levocardia is rare, presenting diagnostic and treatment challenges. This case report discusses a 20-month-old boy with TOF, situs inversus, and levocardia. Initially planned for total surgical correction, intraoperative anatomical complexities led to the decision to perform a bidirectional cavopulmonary shunt surgery. This procedure aimed to enhance oxygen saturation and reduce the volume load on the right ventricle, due to severe pulmonary stenosis and perimembranous ventricular septal defect. The bidirectional cavopulmonary shunt serves to improve systemic arterial oxygen without increasing ventricular work or pulmonary vascular resistance. This case highlights the significance of individualized treatment approaches when TOF is associated with complex anatomical variations.

INTRODUCTION

Congenital heart disease affects approximately 1% of live births, which are typically classified as either acyanotic or cyanotic . Tetralogy of Fallot (TOF) is one of the commonest cyanotic congenital heart defect (Starr, 2010). TOF represents approximately 7% to 10% of all congenital heart disease (Mohammad, 2023).1

Tetralogy of Fallot consists of 4 defects, which are ventricular septal defect, pulmonary stenosis, overriding aorta, and right ventricular hypertrophy (Doyle and Kavanaugh-McHugh, 2019).1 The combination of lesions can be found with a wide spectrum of associate anomalies such as absent pulmonary valve, pulmonary atresia, atrioventricular septal defect, abnormal coronary arteries branching, a right aortic arch and persistence of the left superior vena cava (Ottaviani and Buja, 2022).2

However, its association with situs inversus and levocardia is rarely reported. Scragg and Denny in 1952 reported the first documented case TOF with situs inversus (Sattar *et al.*, 2019). That case was associated with dextrocardia.3 We report a case of these findings in a 20-month-old boy underwent bidirectional cavopulmonary shunt surgery. Congenital heart disease affects approximately 1% of live births, which are typically classified as either acyanotic or cyanotic (Madsen *et al.*, 2016). Tetralogy of

Fallot (TOF) is one of the commonest cyanotic congenital heart defect (Worku and Allen, 2020). TOF represents approximately 7% to 10% of all congenital heart disease (Wald, Valente and Marelli, 2015).1

Tetralogy of Fallot consists of 4 defects, which are ventricular septal defect, pulmonary stenosis, overriding aorta, and right ventricular hypertrophy (Doyle and Kavanaugh-McHugh, 2019). The combination of lesions can be found with a wide spectrum of associate anomalies such as absent pulmonary valve, pulmonary atresia, atrioventricular septal defect, abnormal coronary arteries branching, a right aortic arch and persistence of the left superior vena cava (Ottaviani and Buja, 2022).2

However, its association with situs inversus and levocardia is rarely reported (Marino *et al.*, 2012). Scragg and Denny in 1952 reported the first documented case TOF with situs inversus (Waqar and Riaz, 2016). That case was associated with dextrocardia.3 We report a case of these findings in a 20-month-old boy underwent bidirectional cavopulmonary shunt surgery.

RESEARCH METHODS

This research uses qualitative methods with a literature review approach. Literature review is a systematic, explicit and reproducible method for identifying, evaluating and synthesizing research works and thoughts that have been produced by researchers and practitioners. The step in writing this review literature begins with the selection of topics. Search libraries or sources to gather relevant information from Google Scholar, CINAHL, Proquest, Ebsco, or National Library databases. Determine keywords or keywords for journal searches. After the data is collected, it is processed, analyzed and conclusions drawn.

RESULTS AND DISCUSSION

A 20-month-old boy had been diagnosed with Tetralogy of Fallot at 11 months old and was referred to our hospital with a moderate cyanosis and malnutrition without dyspnea symptoms (Naga *et al.*, 2019). On physical examination, a bluish color of the lips and fingernails was found (Greenberg, Schlosser and Mirowski, 2017). Oxygen saturation level at 88-92%. A grade 3 ejection systolic murmur was also heard at left lower sternal border (Abiko *et al.*, 2019).

Chest x-ray examination showed a cardiomegaly with bronchopneumonia appearance (Lazović, Stajić and Putniković, 2013). An echocardiogram examination showed an overriding aorta (60%), severe infundibular and valvular stenosis, hypoplastic pulmonary valve annulus and large perimembranous ventricular septal defect (Rao, 2022). Cardiac MSCT performed prior to surgery showed the hypertrophy of right ventricle with 10.8 mm thickness, a 14.3-mm-diameter of main pulmonary trunk, an 8.7-mm-diameter right pulmonary artery and a 9.4-mm-diameter

left pulmonary artery. Cardiac MSCT also showed a right sided aortic arch with a 13-mm-diameter of descending aorta.

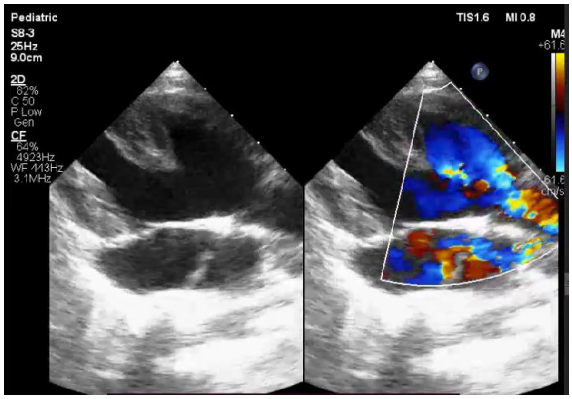

Fig. 1 Preoperative Echocardiogram of Patient

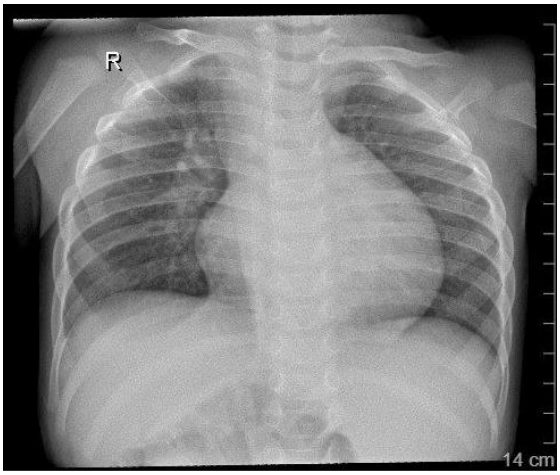

Fig. 2 Preoperative Echocardiogram of Patient

The patient was planed to underwent total correction but during surgical repair, but at the time median sternotomy was performed, intraoperative findings was a left-sided superior vena cava, a left-sided morphological right atrium, left ventricle position was anterior to right ventricle and the aorta is anterior and rightward to the pulmonary artery.

Due to anatomical difficulties it was decided to underwent bidirectional cavopulmonary shunt. Cannulation of the left-sided SVC through left-sided morphological right atrium was performed without cardiopulmonary bypass machine (Kotani *et al.*, 2015). The SVC-pulmonary artery anatomosis was done by 5.0 prolene suture (Al-Amran and Shahkolahi, 2013). After surgery, patient was extubated and monitored in the ICU for twenty hours before moved to general ward. Chest drain was removed on the fifth day and patient was discharged on the seventh day.

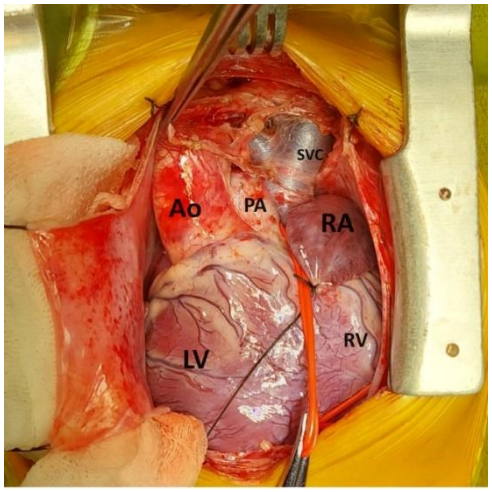

Figure 3. Intraoperative Findings

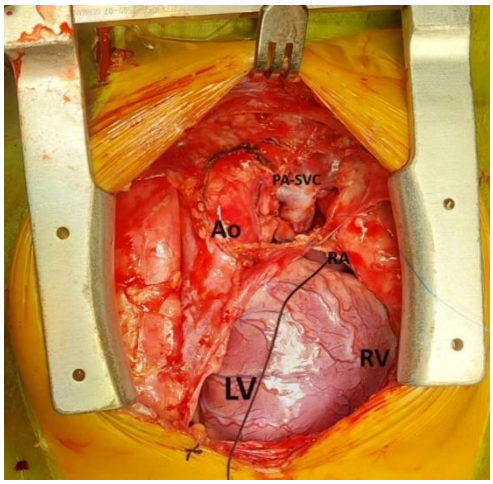

Figure 4. Post Bidirectional Cavopulmonary Shunt Surgery

Tetralogy of Fallot is one of commonest cyanotic congenital heart disease. Until recent decades, surgical correction involved a 2-staged approach: an initial palliative systemic-to-pulmonary arterial shunt, and a later repair to close the VSD and resect the RV outflow muscular culature. Currently, complete single-staged repair is

recommended and earlier at the young age. Although TOF is a common congenital heart disease, its association with situs inversus and levocardia is rare.4

Situs inversus is a rare congenital abnormality condition in wich the major visceral organs are reversed from their normal positions (Gupta *et al.*, 2017). Situs inversus generally occurs with dextrocardia but rarely it may occur with a normally located left sided heart. Various terms have been used to describe this condition such as 'situs inversus with levocardia', 'isolated levocardia' and 'situs inversus incompletus'.3

Situs inversus with levocardia is extremely rare and is almost always associated with congenital heart disease. It has reported incidence of 1:22,000 in the general population and accounts for 0.4-1.2% of all congenital heart diseases. The prognosis is poor, with 5 year survival rate of 5-13% from birth.5

The patient we reported was planned to underwent total surgery correction but at the time median sternotomy was performed, the anatomical difficulties encountered cause total surgery correction cannot be performed. Therefore, this patient uderwent bidirectional cavopulmonary shunt surgery to reduces the volume load on the right ventricle and increase oxygen saturation.

The bidirectional cavopulmonary shunt may be defined as an operation that diverts the systemic venous return from the superior vena cava to both lungs. This surgical connection can provide increased pulmonary blood flow in patients with cyanotic congenital heart disease and reduced pulmonary blood flow.6

The bidirectional cavopulmonary shunt improves systemic arterial oxygen saturation without increasing ventricular work or pulmonary vascular resistance.6 The bidirectional cavopulmonary shunt is undertaken also to enable ventricle volume unloading, associated with a reduction in ventricular end-diastolic volume and ventricular hypertrophy due to volume overload caused by severe pulmonary stenosis and PM-VSD.7

CONCLUSION

Tetralogy of Fallot association with situs inversus and levocardia is a rare case, hence the diagnosis and treatment pose more challenges. Bidirectional cavopulmonary shunt surgery was performed in this patient to increase oxygen saturation and reduces the volume load on the right ventricle due to severe pulmonary stenosis and PM-VSD.

BIBLIOGRAPHY

Abiko, M. *et al.* (2019) 'Cor Triatriatum Dexter in a Infant with a Hypoplastic Right Ventricle', *Journal of Pediatric Cardiology and Cardiac Surgery*, 3(1), pp. 28–34.

Al-Amran, F.G. and Shahkolahi, M.M. (2013) 'Total arterial anastomosis heterotopic heart transplantation model', in *Transplantation Proceedings*. Elsevier, pp. 625–629.

Doyle, T. and Kavanaugh-McHugh, A. (2019) 'Pathophysiology, clinical features, and diagnosis of tetralogy of Fallot', *URL: https://www. uptodate. com/contents/pathophysiology-clinical-features-and-diagnosis-of-tetralogy-of-fallot* [Preprint].

Greenberg, S.A., Schlosser, B.J. and Mirowski, G.W. (2017) 'Diseases of the lips', *Clinics in Dermatology*, 35(5), pp. e1–e14.

Gupta, R. *et al.* (2017) 'Neonatal intestinal obstruction associated with situs inversus totalis: two case reports and a review of the literature', *Journal of Medical Case Reports*, 11(1), pp. 1–6.

Kotani, Y. *et al.* (2015) 'Anatomical risk factors, surgical treatment, and clinical outcomes of left-sided pulmonary vein obstruction in single-ventricle patients', *The Journal of Thoracic and Cardiovascular Surgery*, 149(5), pp. 1332–1338.

Lazović, B., Stajić, Z. and Putniković, B. (2013) 'Rapidly vanishing lung pseudotumor in a patient with acute bilateral bronchopneumonia', *Vojnosanitetski pregled*, 70(9), pp. 878–880.

Madsen, N.L. *et al.* (2016) 'Congenital heart disease with and without cyanotic potential and the long-term risk of diabetes mellitus: a population-based follow-up study', *Journal of the American Heart Association*, 5(7), p. e003076.

Marino, B. *et al.* (2012) 'Abnormalities of situs', *Pediatric Cardiovascular Medicine*, pp. 702–729.

Mohammad, A. (2023) 'Teratology of Fallot (TOF)', in *Clinical and Surgical Aspects of Congenital Heart Diseases: Text and Study Guide*. Springer, pp. 69–85.

Naga, O.I. *et al.* (2019) 'Last-Minute Review', in *Pediatric Board Study Guide: A Last Minute Review*. Springer, pp. 1015–1135.

Ottaviani, G. and Buja, L.M. (2022) 'Congenital heart disease: pathology, natural history, and interventions', in *Cardiovascular pathology*. Elsevier, pp. 223–264.

Rao, P.S. (2022) 'Echocardiography: An overview-part II', *Vessel Plus*, 6, p. 25.

Sattar, Z. *et al.* (2019) 'Tetralogy of Fallot with isolated levocardia in a young female', *Journal of Community Hospital Internal Medicine Perspectives*, 9(3), pp. 275–278.

Starr, J.P. (2010) 'Tetralogy of Fallot: yesterday and today', *World journal of surgery*, 34, pp. 658–668.

Wald, R.M., Valente, A.M. and Marelli, A. (2015) 'Heart failure in adult congenital heart disease: emerging concepts with a focus on tetralogy of Fallot', *Trends in Cardiovascular Medicine*, 25(5), pp. 422–432.

Waqar, T. and Riaz, M.U. (2016) 'TETROLOGY OF FALLOT: WITH DEXTROCARDIA AND SITUS INVERSUS: A CASE REPORT', *The Professional Medical Journal*, 23(10), pp. 1293–1295.

Worku, D. and Allen, R. (2020) 'Tetralogy of Fallot: Origins, management and outcomes', *European Journal of Experimental Biology*, 10(3), p. 10.