Case Report

Role of perioperative transesophageal echocardiography in the management of adolescent truncus arteriosus: Rare case report

P. S. Nagaraja, Naveen G. Singh, Parimala Prasanna Simha, K. R. Davan, V. Manjunath, A. M. Jagadeesh

Department of Cardiac Anaesthesiology, Sri Jayadeva Institute of Cardiovascular Sciences and Research, Bengaluru, Karnataka, India

ABSTRACT

Truncus arteriosus (TA) is a rare congenital heart disease defined as a single arterial vessel arising from the heart that gives origin to the systemic, pulmonary and coronary circulations. The truncal valve in majority of the cases is tricuspid though quadricuspid and bicuspid valves have been reported. Patients with TA typically have a large nonrestrictive sub truncal ventricular septal defect. Survival of these infants beyond 1-year is uncommon. Here, we report a unique case of 12-year-old female patient with persistent TA who underwent surgical repair by using transesophageal echocardiography as a monitoring device during the perioperative management.

Received: 01-11-14
Accepted: 13-02-15

Key words: Pulmonary vascular resistance; Transesophageal echocardiography; Truncus arteriosus

INTRODUCTION

Truncus arteriosus (TA) is a rare congenital heart disease (CHD) with the incidence of 0.03–0.056/1000 live births.[1] Heart has a single arterial outlet with single semilunar truncal valve supplying systemic, pulmonary and coronary circulation. TA patients rarely survive beyond infancy, once diagnosed they would require early surgical repair. Long-term survival is rare.[2] Transesophageal echocardiography (TEE) examination helps in delineating the anatomy of complex CHD before and after surgical repair. Hence we found it worthy to report the importance of TEE examination in the perioperative management of this rare case of adolescent TA with associated right aortic arch, ventricular septal defect (VSD) and incompetent truncal valve.

Access this article online
Website: www.annals.in
DOI: 10.4103/0971-9784.154487
Quick Response Code:

CASE REPORT

A 12-year-old female patient with persistent TA was posted for surgical correction. Her history revealed frequent cough with fever every 3–4 months and hemoptysis since 2 months. On examination, clubbing and cyanosis were present. Parasternal heave was palpable and thrill was present in the left 2nd inter costal space. On auscultation, a single loud S2, systolic murmur in left parasternal area and early diastolic murmur of truncal valve regurgitation was heard. Electrocardiogram showed features of biventricular hypertrophy. Chest X-ray showed features of cardiomegaly and right aortic arch. Cardiac catheterization and angiography showed right aortic arch, normal coronary artery anatomy and pulmonary artery (PA) arising from the posterior aspect of truncus [Figure 1]. The Qp/Qs ratio of 2:1 and an increased pulmonary vascular resistance (PVR) was also noted. Aortic pressure of 131/86 and PA pressure of 127/86 were recorded during cardiac catheterization. Transthoracic echocardiography examination preoperatively reported - CHD with situs

Address for correspondence: Dr. Naveen G. Singh, Department of Cardiac Anaesthesia, Sri Jayadeva Institute of Cardiovascular Sciences and Research, Jayanagar, Bengaluru - 560 069, Karnataka, India. E-mail: naveengsingh@hotmail.com

solitus, PA arising from aorta with a truncal valve over riding interventricular septum (IVS) with moderate truncal valve regurgitation, large subtruncal VSD of 20 mm in size with bi-directional shunt, right aortic arch and normal left ventricle (LV) function.

Intraoperative TEE examination confirmed that the PA was arising posterior to the truncal artery in mid-esophageal (ME) ascending aortic short axis view. The multiplanar angle is slowly rotated from 0° to 50° to clearly delineate the origin of PA from the truncus. By applying color flow Doppler the truncal artery and PA were well visualized [Figure 2a] [Video 1]. A large subtruncal nonrestrictive VSD was visualized in ME aortic long axis view with truncal valve overriding IVS. Moderate truncal valve regurgitation and the VSD shunting bi-directionally was clearly evident on color flow Doppler [Figure 2b]. Patient underwent VSD closure with right ventricle (RV) to PA conduit. Postsurgical repair patient was weaned from cardiopulmonary bypass (CPB) with dopamine of 5 μg/kg/min in sinus rhythm and normal hemodynamics. CPB time was 115 min and aortic cross-clamp time was 88 min.

A detailed TEE examination was performed postsurgical repair. The VSD patch was found to be intact with no residual shunt. Aortic valve (preoperatively truncal valve) showed a minimal regurgitation. Color flow Doppler of RV to PA conduit showed laminar flow with minimal gradient [Figure 3] [Video 2]. Normal biventricular function was noted. Duration of surgery was 5 h and patient was electively ventilated overnight and extubated after 12 h. Dopamine was weaned off in the 1st postoperative day (POD) and patient was shifted to the ward on 2nd POD.

DISCUSSION

Truncus arteriosus, also known as common arterial trunk is a rare CHD (<1%), occurs due to the failure of aortico-pulmonary septum to develop and to separate the embryonic truncus into aorta and main PA.[1] Etiology is mostly genetic, 30-40% due to 22q11.2 microdeletion[3] and rest due to environmental factors such as maternal 1st trimester exposure to alcohol, viral infection, dyes and dietary deficit in vitamins.[4,5]

About 34.8% of TA patients are associated with cardiac and other extracardiac anomalies[6,7] that is, right aortic arch 25–30% cases, interrupted aortic arch, aberrant right subclavian artery, abnormal coronary arteries,

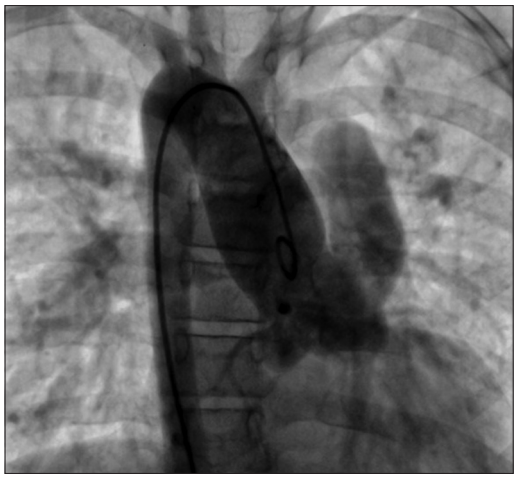

Figure 1: Cardiac catheterization and angiography showing type 1 truncus arteriosus

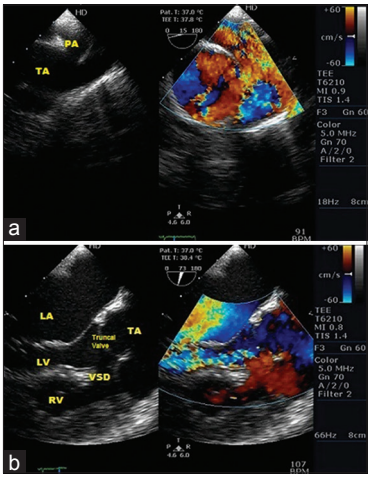

Figure 2: (a) Prerepair mid-esophageal ascending aorta short axis showing type 1 truncus arteriosus. (PA-Pulmonary artery, TA- Truncal artery [truncus arteriosus]) (b) Prerepair mid-esophageal aortic long axis view showing ventricular septal defect and truncal regurgitation (LA - Left atrium, LV - Left ventricle, RV - Right ventricle, VSD - Ventricular septal defect, TA - Truncal artery [truncus arteriosus])

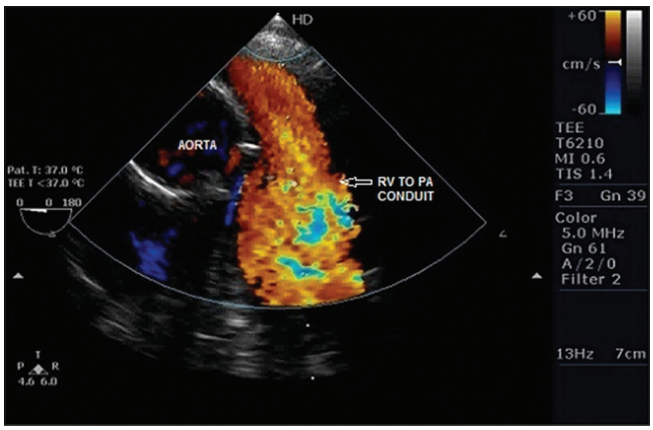

Figure 3: Postrepair mid-esophageal ascending aortic short axis colour flow Doppler showing right ventricle to pulmonary artery conduit

atrial septal defect, tricuspid atresia and double aortic arch.

Truncus arteriosus classification was developed by Collett-Edwards and by Van Praagh[2] based on origin of PA. The patient in this article was Collet-Edwards type 1 and Van Praagh type A1 (single posterior pulmonary trunk).

Truncus arteriosus in infants usually have low PVR resulting in excessive pulmonary blood flow, causing congestive heart failure. Truncal regurgitation if present, can worsen the condition by exaggerating the LV volume status. The reason for survival beyond infancy is mostly due to an increase in the PVR, which relieves the LV volume overload but at the cost of increase in cyanosis.[8] The LV volume overload can also be decreased when there is truncal stenosis or obstruction to the pulmonary blood flow. In our case report, patient had survived till adolescence, probably due to a slowly progressive increase in PVR, which was confirmed by cardiac catheterization. Anesthetic considerations in our patient who had a high PVR was primarily to prevent any factor, which would increase the PVR like hypoxia, hypercarbia and acidosis. Constant hemodynamic monitoring and measures to adjust (particularly) PVR, systemic vascular resistance, and cardiac performance are probably more important than selecting particular anesthetic agent. All anesthetic agents should be carefully titrated to its effect.[2]

The need for intraoperative TEE monitoring in patients undergoing complex CHD surgical repair is increased substantially. Rarely TA patients survive beyond infancy, and our case had survived till adolescence. Since this patient had operable reactive pulmonary vascular physiology, surgical repair was attempted to intervene the natural history of the disease and improve the quality of life. We never wanted this patient to undergo re-exploration for residual VSD shunt or RV to PA gradient, if occured would have resulted in an adverse outcome in postoperative period. Hence, a detailed TEE examination postsurgical repair was performed to interrogate for truncal valve regurgitation, residual VSD, proper functioning of RV to PA conduit and global myocardial function.

CONCLUSION

The complex anatomy of TA in a 12-year-old patient was visualized using TEE in great detail. TEE is an useful tool for perioperative management of patients with TA undergoing surgical repair. Any residual VSD or obstruction to the RV to PA conduit can be identified intraoperatively with TEE, which might be missed otherwise resulting in re-exploration, increased morbidity and increased duration of hospital stay.

REFERENCES

1. Mittal SK, Mangal Y, Kumar S, Yadav RR. Truncus arteriosus type 1: A case report. Indian J Radiol Imaging 2006;16:229-31.
2. McKenzie IM. Truncus arteriosus. In: Lake CL, Booker PD, editors. Paediatric Cardiac Anaesthesia. 4th ed. Philadelphia: Lippincott Williams and Wilkins; 2005. p. 472-9.
3. McElhinney DB, Driscoll DA, Emanuel BS, Goldmuntz E. Chromosome 22q11 deletion in patients with truncus arteriosus. Pediatr Cardiol 2003;24:569-73.
4. Botto LD, Khoury MJ, Mulinare J, Erickson JD. Periconceptional multivitamin use and the occurrence of conotruncal heart defects: Results from a population-based, case-control study. Pediatrics 1996;98:911-7.
5. Tikkanen J, Heinonen OP. Risk factors for conal malformations of the heart. Eur J Epidemiol 1992;8:48-57.
6. Volpe P, Paladini D, Marasini M, Buonadonna AL, Russo MG, Caruso G, *et al.* Common arterial trunk in the fetus: Characteristics, associations, and outcome in a multicentre series of 23 cases. Heart 2003;89:1437-41.
7. Ferdman B, Singh G. Persistent truncus arteriosus. Curr Treat Options Cardiovasc Med 2003;5:429-438.
8. Perloff JK. Truncus arteriosus. In: Perloff JK, editor. Clinical Recognition of Congenital Heart Disease. 5th ed. Philadelphia: W. B Saunders; 2003. p. 541.

Cite this article as: Nagaraja PS, Singh NG, Simha PP, Davan KR, Manjunath V, Jagadeesh AM. Role of perioperative transesophageal echocardiography in the management of adolescent truncus arteriosus: Rare case report. Ann Card Anaesth 2015;18:234-6.

Source of Support: Nil, **Conflict of Interest:** None declared.